X-ref *For other Roundups in this issue that cross-reference with Oncology see: Spine Roundup 7; Research Roundup 1.*

Intralesional treatment in low-grade chondrosarcoma

Low-grade chondrosarcoma (LGCS) has traditionally been treated with wide local resection, with the aim of clear margins while maintaining and preserving function. More recently, there has been a growing movement towards intralesional surgery, with its accompanying improved function and reduced complication rates. However, without evidence that survival is not compromised, pursuing this technique could be storing up a lot of problems in the future. A group from **Groningen (The Netherlands)** have investigated intralesional curettage of central long bone LGCS, using a systematic review of retrospective studies and case series taken from the usual electronic biomedical databases.[1] The aim of their review was to evaluate the recurrence-free survival, tumour upgrading, complications, and functional outcome of patients treated with intralesional curettage when compared with wide local excision as the benchmark. Of the 511 participants included in the analysis, 419 were treated with intralesional treatment and 92 were treated with wide local resection. Meta-analysis failed to show any significant differences in recurrence-free survival after intralesional treatment *versus* wide resection, although the functional scores were better after intralesional treatment compared with wide resection. This was also reflected in morbidity, with lower reported complication rates in patients treated with intralesional surgery. In a result that swung the other way, Kaplan–Meier analysis showed a rate of 96% recurrence-free survival after resection *versus* 94% following intralesional treatment. However, given the relatively small number of patients in this analysis, it is not really possible to draw inferences from a 2% difference in survival. Despite an extensive literature search, the authors conclude that, while intralesional treatment is common and apparently successful for LGCS, there are no reliable prospective randomized controlled trials on the subject. In the absence of large cancer registry studies, evidence synthesis based on case series is the best current available evidence, as presented here. Furthermore, there remains the ever-present diagnostic problem that the true criteria to distinguish a benign 'atypical cartilage tumour' from a LGCS has yet to be clarified.

Locally recurrent chondrosarcoma of the pelvis and limbs: treatment options

One of the most feared outcomes following treatment for bone tumours is recurrence, and local recurrence in the pelvis is particularly bad news for patients and surgeons alike. Local recurrence in the pelvis is often late to present due to the lack of local symptoms, and is located in an area that is incredibly difficult to access. Treatment options for revision surgery in the limbs and pelvis can, therefore, be limited and technically challenging. The anatomical challenges are added to by scarring and poor tissue stock, and so a thorough understanding of the precise nature of the surgical treatment of recurrent tumours is essential. In this study from **Birmingham (UK)**, the authors sought to investigate the factors that influence the successful surgical control of recurrent chondrosarcoma of the pelvis or lower limb.[2] Using a departmental registry from a regional referral database, the authors identified 126 patients with local recurrence of a chondrosarcoma of either the pelvis or lower limb between 1990 and 2015, sourcing records of two years' follow-up and the histopathology for each. The study group of 126 patients represented a local recurrence rate of around a quarter. There was adequate prospectively collected clinical data, including anatomical details of the tumour, surgical details, margin achieved, and patient demographic factors. Outcomes available included local recurrence-free survival and disease-specific survival. All of these factors were analyzed using univariable and multivariable analysis to establish the likelihood of recurrence and the best treatment options. The authors found that in those patients without metastases prior to – or at the time of – the diagnosis of a recurrence, disease-specific survival was 62.5% at one year and 45.5% at five years. Grade, surgical margin, age at time of recurrence, interval between primary surgery, and recurrence were significant risk factors associated with survival. Wide surgical margins were clearly associated with a reduced rate of subsequent recurrences compared with marginal margins or intralesional treatment. Overall, this study shows that surgery remains the best treatment for chondrosarcoma, and that clear margins are essential for optimum outcome both with primary tumour and recurrence.

Is limb-sparing surgery comparable to amputation in pelvic chondrosarcoma?

Broadly speaking, there are two surgical strategies open to a surgeon operating on musculoskeletal tumours: limb-sparing surgery and amputation. With progress being made both in adjuvant treatments and in surgical care and techniques, there is a trend towards limb-sparing surgery in the management of pelvic chondrosarcoma, one of the more common tumours of bone. However, evidence is currently lacking as to whether this leads to improved long-term survival. Addressing this question, a group from **Cleveland, Ohio (USA)** have used the National Cancer Database to identify 516 patients undergoing either amputation or limb-sparing treatment for pelvic chondrosarcoma. The authors created two matched cohorts of 131 patients using propensity score matching.[3] They found no difference in survival between limb sparing and amputation, which at five years was 70% in each category, and at ten years was 60%. Older age, higher grade, and increased comorbidities were associated with decreased survival, and yet again positive margins were shown overall to be a poor prognostic indicator. On the face of it, this study seems to demonstrate that limb salvage is as safe as amputation for pelvic chondrosarcoma. The authors comment that the relevance of case selection cannot be ruled out, and that it is likely that outwardly more aggressive-appearing tumours were selected for amputation. As a result, the fact that the survival for the two groups was identical is encouraging.

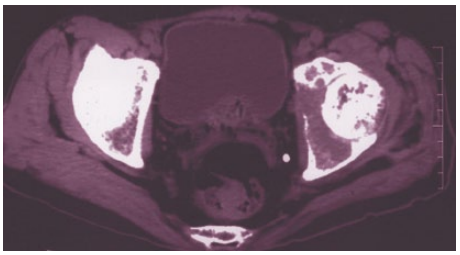
Ewing's sarcoma diagnosis: MRI or scintigraphy?

The staging of tumours forms an important part of the early management of bone lesions. There are a number of considerations in imaging, not just the sensitivity and specificity from a diagnostic perspective. In modern times, with new techniques arriving all the time, a group from **Stanmore (UK)** has collected and reported their experience of imaging investigations in patients with a diagnosis of Ewing's sarcoma.[4] The aim of the study was to compare the diagnostic accuracy of staging of these lesions with either whole-body MRI or scintigraphy. A total of 180 paediatric and adult patients were identified as part of the review between 2007 and 2018, with skeletal metastases identified in 30. A total of 96 patients underwent MRI, with metastases found in 23, and 118

underwent scintigraphy, with metastases found in 20. In all, 70 patients were investigated with both modalities, with 13 patients having metastases identified in both MRI and scintigraphy. Four patients showed the presence of metastases in MRI alone, while none were found in scintigraphy alone. The authors specifically note that scintigraphy superseded MRI in detecting skull vault metastases, and correspondingly showed false positives in long bone physes. This paper shows impressive results, suggesting that bone scans have been superseded by whole-body MRI for staging of Ewing's sarcoma. It remains to be seen if these results can be translated to other tumours. This finding is particularly important in paediatric tumours, where the doses of radiation associated with whole body scintography are not insignificant and are known to be harmful to the skeletally immature.

Does timing affect survival in Ewing's sarcoma?

■ Many factors likely influence the success of treatment in Ewing's sarcoma. The regime and its modality are certainly key, but timing of therapy could also have a role in influencing successful treatment. A group from **Houston, Texas (USA)** have investigated this topic using the National Cancer Database.[5] The authors identified 1318 patients with Ewing's sarcoma. All of the patients were treated with primary chemotherapy and subsequent local chemotherapy. The study cohort were analyzed with a series of Kaplan–Meier survival curves. The authors then went on to use multiple binomial logistic regression to identify factors linked with a prolonged delay to local therapy. This was further analyzed with a Cox hazards model to find those factors relating to overall survival. Their analyses demonstrated that being older than 21 years of age, having a tumour size greater than 8 cm, a time to local chemotherapy of over 16 weeks, and positive margins following excision were associated with a reduced overall survival. The rise of 'big data' and national registries as almost the default method for investing epidemiological and treatment questions certainly has its problems. There are a number of detractors who argue that these datasets are designed for, and only appropriate for, monitoring efficacy of treatments and performance of centres and individual clinicians. While there is certainly some merit in the arguments put forward from this perspective, studies like this for a rare diagnosis clearly could not be performed in another way. This helpful study confirms the importance of early local therapy, whatever its modality over delayed treatment, and again shows that negative margins are a must with surgical treatment.



Desmoid tumours: excise or observe?

■ The traditional treatment for patients presenting with desmoid tumours has been in the hands of the surgeon, with excision being the mainstay of care. However, these are, for the most part, indolent connective tissue tumours. As time has passed, it has increasingly become a condition treated conservatively with good results reported, and occasionally with the addition of drugs to accelerate recovery. Despite this shift in 'standard care', there are few studies that support treatment in one way or another. In this study from **Calgary (Canada)**, the local surgical oncology research group reported on 111 patients previously treated in their centre, and aimed to establish the expected success rate with simple observation alone.[6] The group identified their cohort from the Alberta Cancer Registry, using patients diagnosed between 2004 and 2015. Outcomes of immediate surgery and conservative waiting were recorded. In all, 74% of the identified patients were female, and median follow-up was 35 months. Overall, 45% of patients were treated conservatively, and disease progression was observed in 42% of these. The remainder were treated with resection at presentation, 15% of which showed recurrence. Interestingly, the microscopic margin had no effect on the risk of recurrence after surgery. Overall, observation was the definitive successful treatment in 58% of patients. Desmoid tumour is fast becoming a nonsurgical disease, although surgery had the more favourable reported rate of recurrence. As ever, the authors recommend a multidisciplinary approach, but, based on the results presented here, almost two-thirds of patients will need nothing more than a period of watchful waiting.

Denosumab and giant cell tumours of bone

■ Giant cell tumours of bone (GCTB) are unusual, but can present with catastrophic symptoms warranting urgent treatment. Those of us who do not see tumours daily might be less familiar with the developments in managing these lesions. Denosumab, a monoclonal antibody that targets the RANK ligand, has been approved for use in skeletally mature patients and has dramatically changed the treatment landscape of GCTB. There has been a raft of papers to support its use, many of which have been previously covered in *360*. Despite the undoubted successes of denosumab as therapy for GCTBs, the initial enthusiasm has been tempered by caution, and the best use of this new systemic treatment modality is currently the subject of much discussion. There have been concerns raised surrounding the recurrence once therapy is ceased, as well as around the side-effect profile of the treatment itself. Fortunately, a group from **Leiden (The Netherlands)** have summarized the challenges of GCTB treatment with denosumab and have reviewed recent publications and new insights on this subject.[7] Overall, the authors paint a positive picture. Using neoadjuvant denosumab appears to downstage tumours, facilitating surgical treatment, halting tumour progression when unresectable, and improving symptoms. On the other hand, the team note that concerns exist about local recurrence following surgery, that osteonecrosis of the jaw remains a risk, and that the optimal treatment scheduling and duration are as yet undefined. Denosumab is still in the investigative phase of its use in GCTB; however, current evidence is promising and is, based on existing studies, likely to positively influence the care of patients with GCTB. We would recommend this paper for any surgeon preparing for exams, or for those involved in the decision making and treatment processes surrounding giant cell tumours.

The Ilizarov method and bone defects in tumour

■ The application of bone transport using the Ilizarov method for patients diagnosed with osteosarcoma following surgical resection is uncommon, largely because of the perceived disadvantages of pin-tract infection, refracture, and poor and slow bone formation in patients receiving chemotherapy. There are also concerns about pin tracts and regenerate quality in any limb following radiotherapy. However, given the difficulty of bone defects in tumour patients and the ongoing difficulties with reconstruction following limb-salvage procedures, a group of authors from **Beijing (China)** have documented the results of ten patients with limb osteosarcoma receiving limb-salvage treatment by the Ilizarov method and distraction osteogenesis.[8] The transported fragment of bone had a mean length of 14 cm in this series, and was transported for a mean of 140 mm. Patients spent an average of 69 months in their circular frame, and

seven patients required further surgery to treat postoperative complications. The group found that ten months after fixator removal, both the Association for the Study and Application of the Method of Ilizarov (ASAMI) bone score and ASAMI function score were excellent and good in 80% and 20% of cases, respectively. These results are similar to those expected from traditional bone transport for infection or other diagnoses. The authors conclude that despite well founded theoretical reservations, these did not translate into clinical practice. Undertaking bone transport using the Ilizarov method can achieve good outcomes following limb salvage in patients treated for osteosarcoma with neoadjuvant chemotherapy, provided that complications are recognized early and are managed confidently and precisely.

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Children's orthopaedics

X-ref ***For other Roundups in this issue that cross-reference with Children's orthopaedics see: Oncology Roundups 4 & 5.***

The Dunn procedure in SCFE

The modified Dunn procedure has been utilized for many years to restore the anatomy of hips with severe slipped capital femoral epiphysis (SCFE). The progression of SCFE can lead to a severe deformity, which in turn can lead to femeroacetabular impingement and early osteoarthritis. The logic behind the application of the modified Dunn procedure is the desire to correct the anatomy to ameliorate this risk. This, of course, somewhat simplifies the decision making where the main operative risk is of avascular necrosis (AVN) due to interruption of the blood supply to the femoral head at the time of surgical correction. From a population perspective, the longer-term outcomes are a balance in the increased risks of AVN due to operative intervention and the increased risk of osteoarthritis from femoroacetabular impingement if malunion occurs. There has been increased interest in the Dunn osteotomy in recent years as the link between impingement and osteoarthritis has become more firmly established. While previous series from this group from **Bern (Switzerland)** have included all slip angles, the current paper examines exclusively the unit's experience of severe slips with a slip angle greater than 60°.[1] The review is retrospective, but the unit's entire experience of 131 hips treated with a modified Dunn procedure from 1999 to 2016 was reviewed and 46 hips with a severe slip angle were included. At nine-year follow-up, 40 hips were available for clinical and radiological review. In terms of the development of osteoarthritis, one hip showed progression, and the group had a mean Hip Disability and Osteoarthritis Outcome Score of 91 points. However, the follow-up here is relatively short as these patients are now only in their third decade. Functional hip scores were high, with a mean Harris Hip Score of 94 points. Importantly, two patients developed AVN and underwent further surgery, but the authors do state that these patients both had acute-on-chronic slips and were unstable intraoperatively, even if considered stable by the Loder classification. Three patients underwent revision due to breakage of screws or change of wires. Overall, the cumulative survival was 86% at ten-year follow-up. This series does seem to support recent changes in practice towards operative intervention. Based on these results, in expert hands and in a unit with large experience, this seems a reliable option to normalize the anatomy in what is a difficult problem to treat. The occurrence of AVN is still a concern and MRI was not used to detect this here, so only disease visible on plain film was included in the results. However, this may be clinically appropriate, especially given the relatively long follow-up interval for which any AVN-related collapse could be expected to be seen. We look forward to longer-term results from this series, particularly with respect to osteoarthritic disease.

What is the cartilage pressure in the Pavlik harness?

Pavlik harness treatment is perhaps the most commonly employed means of treating developmental dysplasia of the hip (DDH), but our understanding of the changes we are introducing to the native hip environment is limited. The Pavlik harness works to constrain the hip joint, thereby reducing the eventual instability and promoting normal development. The downside to this approach is the risk that increased cartilage contact pressures may impede nutrition to the cartilage, or that the pressure may directly damage the ossific nucleus or cartilage. In this study from **Edmonton (Canada)**, the authors utilized finite element modelling of normal and dysplastic hips in a Pavlik harness to look at the internal loading environment of the hip in terms of cartilage contact pressures (CCP).[2] In most clinical cases, our ignorance of these pressures is perhaps not relevant; however, in difficult hips, or in those patients who develop avascular necrosis, it is instructive to consider what the contact pressures might be when there are a range of treatment options available. This group therefore developed a finite element model of CCPs in cartilaginous infant hips subjected to Pavlik harness treatment. Normal and dysplastic hips in a Pavlik harness at 90° flexion and gravity-induced abduction angles of 40°, 60°, and 80° were modelled. In the harness, the contact pressures were found to be distributed in a horseshoe shape along the anterior and posterior margins of the acetabulum, leaving the roof relatively lightly loaded in the normal hip model, but unloaded in the dysplastic hip; it is these pressures that are thought most likely to stimulate the remodelling process. At 40° of abduction in the dysplastic hip, the model predicted equilibrium in a dislocated position, which is