

For other Roundups in this issue that cross-reference with Oncology see: [Paeds Roundup 5, 7](#); [Research Roundup 3, 4](#).

## Radiotherapy for the radioresistant

■ Chordoma and chondrosarcoma were, until recently, considered to be radioresistant tumours and the only treatment option was surgical. Newer protocols for high-dose local radiotherapy have proven to be effective and chordomas are now often treated with high-dose radiotherapy. Researchers in **Osaka (Japan)**, taking note of early evidence that chondrosarcomas may also be treated in this manner, have reported their experience of treatment of large chondrosarcomas in the pelvis with either surgery or carbon ion radiotherapy (CIRT). This is a particularly relevant concept for large pelvic chondrosarcomas where conventional treatment is usually administered with debilitating surgery, such as a hindquarter amputation. The report is of a comparative series of 31 patients, all with chondrosarcoma of the pelvis treated with either surgical resection or CIRT. It was a fairly mismatched series, with just seven patients receiving the CIRT due to the retrospective nature of the study. The treatment allocation did not affect overall survival (72% five-year survival) or indeed recurrence rates (32% local recurrence rate).<sup>1</sup> Proton radiotherapy will be coming to the UK in a few years and may find a role in helping treat these difficult tumours, especially given the better functional outcomes reported in the CIRT group in this study.

## Multiple hereditary exostosis

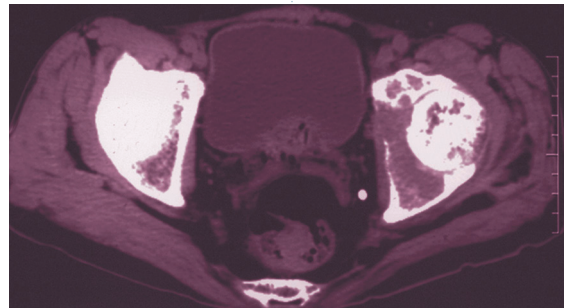
■ Hereditary multiple exostosis (HME) is an autosomal dominant condition which results in large numbers of skeletal exostosis. Patients undergo lifelong surveillance as the malignant transformation rate is known to be high. There are, however, few large-scale studies, and

to date the disease progress, risk of malignant transformation and incidence data are based on rather small population studies. In a very simple but to-the-point study, researchers from **New York (USA)** undertook a web-based survey distributed via online 'disease support groups'. The survey included relatively valuable data on disease transformation in the 779 respondents, estimating the risk of malignancy in HME and serving a useful purpose in informing patients.<sup>2</sup> Given the lifetime risk of anyone getting cancer is at around 50%, this puts the 2.7% risk reported here in perspective.

## The total femur as a limb salvage option

### x-ref Hip, Knee

■ In advanced femoral oncological disease the only prospect for limb salvage can be a total femoral arthro-



plasty. These large prostheses are known to have significant infection rates and high morbidity. However, there are few papers describing their outcomes in the longer term, and establishing the clinical outcomes in terms of function and survival has been limited by the lack of a large study. Oncology surgeons in **Vienna (Austria)** describe their own large series of total femur arthroplasties. The study cohort consisted of 50 patients undergoing total femur replacement with resection for localised primary bone tumours. The series reports a lengthy follow-up period of 57 months for conventional prostheses, and 172 months for expandable prostheses.

The conventional implants did better than the expandables, with a five-year survival of 48% versus 30%, respectively.<sup>3</sup> This paper paints an accurate picture of total femur arthroplasty and describes a high morbidity procedure, but the outcomes will still be better than with amputation.

## Survival prediction in osteosarcoma

■ In many branches of oncology the use of interactive survival models (based on nomograms) are central to the assessment and treatment of patients with a variety of cancer subtypes. Currently, however, there is no widely used nomogram to aid decision making in osteosarcoma. In an impressive study, researchers in **Tokyo (Japan)** set out to develop and validate nomograms to predict the survival of patients

with osteosarcoma. Using prospective data from 1070 patients treated with neo-adjuvant chemotherapy for non-metastatic osteosarcoma, multivariate nomograms were developed based on six pathological variables. This large retrospective study confirms that the most important prognostic factors for osteosarcoma are site (proximal in limb versus distal), patient age and response to chemotherapy. Other factors are less important than these three. Interestingly, these authors found that tumour size was no longer a good prognostic factor in this cohort.<sup>4</sup> The next stage in augmented decision making would be interactive nomograms where other tumour factors

such as chemotherapy type, regime and metastasis can be included to guide treatment decisions as well as prognosis.

## What happens when chondrosarcoma recurs?

■ To say that the literature on the outcomes following local recurrence (LR) of chondrosarcoma is scarce is a monumental understatement. There are very few papers available to guide treatment or evidence for prognosis.<sup>5</sup> The medical team in **Seoul (South Korea)** have stepped up to the mark again and assembled a remarkable 28 patients with locally recurrent chondrosarcoma from their total cohort of 150 chondrosarcoma patients. The study team concluded that long-term survival of locally recurrent chondrosarcoma is achievable in a substantial number of patients (post-LR survival at five years was 59%). In terms of predictors of poor outcomes, age greater than 50 years and local recurrence within one year of primary surgery independently predicted poor survival. The authors also manage to reach the rather obvious conclusion that wide surgical margins at revision surgery reduces the risk of subsequent local recurrence. Their observations also led them to suggest that higher rate of LR in axial tumours is not only related to the higher rate of inadequate margins, but also to the more aggressive biology. Interestingly, progression of grade at local recurrence was noted in only one case in their series, whereas reports in the literature suggest a range of between 10% and 35% progression of grade at local recurrence.

## Thumbs up for vascularised fibular graft

■ There will never be a 'best' limb reconstruction option, as the decision making is complex and different for each patient. One of the advantages of vascularised fibular graft (VFG) is that once incorporated, the graft is



'live' and should be resilient and long-lasting, at the trade-off of potentially high initial complication rates and technically demanding surgery. This study from **Leiden (The Netherlands)** describes the outcomes of 74 consecutive patients from four tertiary centres for orthopaedic oncology, all of whom underwent reconstruction using a VFG after resection of a tumour between 1996 and 2011. There were 52 primary and 22 secondary reconstructions, with an impressive 93% (n = 69/74) of patients having successful limb salvage; all of these united and 65 (88%) showed hypertrophy of the graft. In around half of patients (n = 35/74), at least one complication arose, with a greater proportion in lower limb reconstructions, non-bridging osteosynthesis, and in children. Union was not markedly influenced by the need for chemo or radiotherapy, but should not be expected during chemotherapy.<sup>6</sup> The important lessons from this detailed study include that the use of bridging plate osteosynthesis is associated with a reduction in the risk of fracture and the need to restrict weight-bearing for at least 12 weeks post-operatively, especially in children and in the presence of neo-adjuvant treatment.

### Radiotherapy and survival

■ There is a global consensus that external beam radiation therapy (RT) is indicated for all large, deep, high-grade soft-tissue sarcomas (STS). The vast majority of studies have investigated the impact of radiation using local control as an endpoint, rather than overall survival. An epidemiology team from **Taipei (Taiwan)** utilised data from the National Cancer Database (NCDB) to establish what proportion of patients with STS undergo external beam RT.<sup>7</sup> They were able to include the results of 10 290 patients and found that a total of 3982 (37.8%) did not receive RT. This large database review reveals a striking lack of utilisation of RT to treat high-grade STS, which correlated with poorer survival. Lower education and income levels and diminished access to medical care (insurance and distance to the facility) were also associated with failing to receive RT. Unfortunately, the NCDB did not include information regarding local recurrence rates. Therefore, the authors were not able to assess whether the group with increased RT use had higher rates of local control, which may have led to a small, but statistically significant, improved survival (1.2% vs 0.2%) in such a large sample size. They have, however,

suggested that it was conceivable that RT may have led to improved survival independent of its defined role in improving local control, as there are emerging data that radiation therapy can stimulate the immune system, which could theoretically attack distant disease. A study with more data on outcomes is clearly needed here to establish exactly what's going on.

### Musculoskeletal tumours in pregnancy

■ We would finally draw the attention of the 360 readership to a good overview from **Munich (Germany)** of what is an uncommon situation that clinicians may face, that of musculoskeletal tumours in pregnancy, where it is a challenge to make the right decisions at the right time.<sup>8</sup> The main message of the review is the diagnostic difficulties facing surgeons in the diagnosis of a growing painful mass in the pregnant woman. Early diagnosis and careful follow-up at a specialist tertiary referral centre is a necessary doctrine in the management of this complex problem.

### REFERENCES

1. **Outani H, Hamada K, Imura Y, et al.** Comparison of clinical and functional outcome between surgical treatment and carbon ion

radiotherapy for pelvic chondrosarcoma. *Int J Clin Oncol* 2015;(Epub ahead of print) PMID: 26150259.

2. **Czajka CM, DiCaprio MR.** What is the proportion of patients with multiple hereditary exostoses who undergo malignant degeneration? *Clin Orthop Relat Res* 2015;473:2355-2361.

3. **Sevela F, Schuh R, Hofstaetter JG, et al.** Total femur replacement after tumor resection: limb salvage usually achieved but complications and failures are common. *Clin Orthop Relat Res* 2015;473:2079-2087.

4. **Ogura K, Fujiwara T, Yasunaga H, et al.** Development and external validation of nomograms predicting distant metastases and overall survival after neoadjuvant chemotherapy and surgery for patients with nonmetastatic osteosarcoma: A multi-institutional study. *Cancer* 2015;(Epub ahead of print) PMID: 26194185.

5. **Kim HS, Bindiganavile SS, Han I.** Oncologic outcome after local recurrence of chondrosarcoma: Analysis of prognostic factors. *J Surg Oncol* 2015;111:957-961.

6. **Hilven PH, Bayliss L, Cosker T, et al.** The vascularised fibular graft for limb salvage after bone tumour surgery: a multicentre study. *Bone Joint J* 2015;97-B:853-861.

7. **Hou CH, Lazarides AL, Speicher PJ, et al.** The use of radiation therapy in localized high-grade soft tissue sarcoma and potential impact on survival. *Ann Surg Oncol* 2015;22:2831-2838.

8. **Postl LK, Gragl G, von Eisenhart-Rothe R, et al.** Management of musculoskeletal tumors during pregnancy: a retrospective study. *BMC Womens Health* 2015;15:48.

## Children's orthopaedics

**For other Roundups in this issue that cross-reference with Children's orthopaedics see: Trauma Roundup 5, 6; Research Roundup 3, 4.**

### Radiographic follow-up of DDH

■ It is common practice in many institutions to undertake at least some follow-up of patients with a previous history of developmental dysplasia of the hip (DDH), even if the patient has returned to 'normal' on ultrasound scan. The authors of this study in **Philadelphia (USA)**

examine whether this is really strictly necessary. They report a consecutive series of 115 patients with idiopathic DDH presenting to their institution over four years. All of these patients had undergone a clinical and ultrasonic examination within normal limits at a mean follow-up of 3.1 months. Perhaps surprisingly in their series, by age six months, 17% demonstrated radiographic signs of acetabular dysplasia. No significant differences were evident in the six- or 12-month rate of dysplasia between the infants successfully treated with

a Pavlik harness and those infants normalising without treatment, but with a history of risk factors.<sup>1</sup> The conclusions that can be drawn from this paper are very important, in that the paper demonstrates that there is a notable incidence of radiographic dysplasia after apparent normalisation at three months. The study suggests that the risk of radiation exposure is outweighed by the risk of silent and unrecognised dysplasia. The recommendation made by the authors is that radiographic follow-up in this population should

continue at least until walking age, to allow the timely diagnosis of residual acetabular dysplasia. This is a very simple message that has been elegantly demonstrated in a single-centre review, with clear entry and exclusion criteria.

### When the supracondylar goes wrong

#### x-ref Trauma, Shoulder & Elbow

■ Even experienced orthopaedic surgeons will admit to a slight personal tachycardia when a pulseless supracondylar fracture is admitted through the emergency department.

