Table 9. Primary tumors. Surgical treatment. General principles

We recommend:

1.	**en bloc resection in cases of benign aggressive tumors of Enneking stage 3 (i.e., osteoblastomas and giant cell tumors) and low-grade malignant tumors of Enneking stage I A and B like chordomas and chondrosarcomas.**	C	[7]	V	Expert opinion
2.	**the Weinstein Boriani Biagnini staging system as helpful in surgical planning of en bloc resection.**	C	[8, 9]	V	Expert opinion

Table 10. Sarcomas. Treatment. General principles

We recommend:

1.	**combination therapy within established therapeutic protocols for all of the Ewing group's sarcomas and bone sarcomas.** These tumors are highly malignant neoplasms. The results of surgical treatment alone are poor with less than 20% of 5-year survival.	A	[37-50]	I	Randomized control study
2.	**that surgical treatment of Ewing sarcoma be preceded by chemotherapy.**	A	[37-50]	I	Randomized control study

3.	**high dose adjuvant photon/proton radiotherapy of chordomas, chondrosarcomas, and other sarcomas** This kind of radiotherapy provides high local control while late morbidity appears to be acceptable.	B	[51]	III	Prospective
4.	**that surgical treatment be aimed at cure rather than palliation whenever possible**	C	[52]	III	Retrospective comparative study
5.	**periosteal osteosarcoma as the only exception to the use of perioperative chemotherapy is.**	C	[53]	IV	Retrospective

Table 11. Primary malignant tumors. Surgical management

We recommend:

1.	**Enneking marginal and wide resections (Enneking appropriate resections) over the intralesional resection** Enneking marginal and wide resections result in a lower risk of recurrence at the operated site and longer survival than the "intralesional" excision according to Enneking. Favorable oncological outcomes after en bloc resection may be achieved in terms of recurrence and survival. “Intralesional” procedure (when the surgeon incidentally or intentionally violates margins of the tumor) worsens the prognosis and recurrent rate	B/C	[54-56]	II/III	Multicenter ambispective cohort analysis, prospective cohort study, retrospective review
6.	**multilevel en bloc spondylectomy by experienced** Oncologic resections achieved by multilevel en bloc spondylectomy in experienced hands can lead led to an acceptable survival rate with reasonable local control.	C	[57]	IV	Case series

Table 12. Sarcomas. Surgical treatment. General principles.

We recommend:

1.	**en bloc resection rather than piecemeal resection even if both have negative margins.**	C	[52]	III	Retrospective comparative study

	En block resection with tumor-free margins has a lower rate of recurrence than a piecemeal resection with negative margins				

Table 13. Chondrosarcoma. Adjuvant and stand alone radiotherapy

We recommend:

1.	**adjuvant radiotherapy after complete resection of tumour.** Although surgery with complete resection is paramount in management of chondrosarcoma, RT is a useful adjuvant treatment and appears to offer excellent and durable local control where wide surgical resection is difficult to accomplish.	B	[58]	III	Retrospective
2.	**whenever possible high-dose proton irradiation rather conventional radiotherapy after maximum resection of the tumor.** Maximum surgical resection followed by high-dose proton irradiation results in superior results compared with conventional x-ray treatment of chondrosarcomas of the skull base	B	[59-61]	III	Retrospective
3.	**irradiation in case of inoperable tumors.** There is evidence to suggest chondrosarcoma is not radioresistant and irradiation should be considered when surgery would cause major unacceptable morbidity or be technically impossible. Early evidence suggests there may be a role for chemotherapy to supplement the effects of irradiation	B	[62, 63]	III	Retrospective

Table 14. Osteosarcoma.

We recommend:

1.	**Enneking appropriate en bloc (EA) rather than Enneking inappropriate (EI) intralesional resection of the tumor.** There is a significant decrease in recurrence, an increase in survival rate months and a lower metastases development with EA en bloc resection when compared with EI intralesional resection.	B/C	[64-65]	III	Ambispective cohort studies, systematic review with metaanalysis
2.	**high dose proton therapy doses for some patients with unresectable or incompletely resected osteosarcomas.** Proton therapy to deliver high radiotherapy allows locally curative treatment for some patients with inoperable tumour or tumors partially resected.	B	[66-68]	III	Retrospective, Case control study
3.	**radiotherapy or proton therapy after surgical treatment with positive margins.** Radiotherapy can help provide local control of osteosarcoma for patients in whom surgical resection with widely, negative margins is not possible. It appears to be more effective in situations in which microscopic or minimal residual disease is being treated.		[69]	III	Retrospective
4.	**discussion with medical oncologist about adjuvant or neoadjuvant chemotherapy.** The effect of adjuvant and neoadjuvant chemotherapeutics requires further exploration	B	[64]	III	Ambispective cohort studies

Table 15. Ewing sarcoma

We recommend:

1.	**neoadjuvant chemotherapy.** Preoperative chemotherapy allows for satisfactory results in terms of relapse-free survival (RFS). Patients with resectable tumors after initial chemotherapy have a low local failure rate. Some studies show however that with preoperative radiotherapy local control is comparable to that with preoperative (neoadjuvant chemotherapy). The results of axial tumors treatment are comparable to that of appendicular tumors	A	[45, 50, 70]	I	Randomized control study, Retrospective analysis of randomized control studies

2.	**surgical resection whenever possible and appropriate.** Compared to stand-alone radiotherapy for locally advanced disease, a surgical procedure allows for better results in terms of local control (LC) but with no difference in overall survival (OS). Risk of local failure is greater for stand-alone radiation compared to surgery.	A	[49]	I	Retrospective analysis of randomized control studies
3.	**postoperative radiotherapy after intralesional or marginal resections and after wide resection with a poor histologic response.** Postoperative radiotherapy may improve local control after resections with positive margins or even Enneking wide resections in patients with poor histologic response.	A	[45, 71, 72]	I	Retrospective analysis of randomized control studies
4.	**radiotherapy in inoperable cases.**	A	[50]	I	Retrospective analysis of randomized control studies

Table 16. Chordoma

We recommend:

1.	**Enneking appropriate resection.** Enneking appropriate resection plays a major role in decreasing the risk for local recurrence in patients with chordoma of the mobile spine	B	[73]	II	multiinstitutional retrospective study

2.	**postoperative proton therapy rather conventional therapy** Postoperative proton therapy has better overall survival results compared to postoperative conventional photon radiotherapy. 10-year overall survival is higher also for proton therapy than for stereotactic radiotherapy.	B	[74, 75]	II	Metaanalysis, retrospective
3.	**aggressive therapy combining a resection as radical as possible with postoperative proton or radiotherapy.** A combination of aggressive surgery and radiotherapy seems to improve the prognoses of suboccipital and cervical chordomas when applied at the patient's first presentation with the disease. Postoperative radiotherapy gives better survival results than salvage radiotherapy in local recurrence.	B	[76]	III	retrospective
4.	**high-dose definitive radiation therapy in inoperable cases.** In certain circumstances where resection of mobile spine or sacral chordoma may result in significant neurologic or organ dysfunction, patients can be treated definitively with the use of high-dose definitive radiation therapy	B	[77]	III	retrospective

Table 17. Osteoblastoma

We recommend:

1.	**total excision whenever possible.**	B	[28]	III	retrospective
2.	**en bloc resection of stage 3 tumours and allow for intralesional excision of stage 2 lesions.** Total resection is important as local recurrence was found to be strongly associated with mortality. Subtotal excision together with higher preoperative alkaline phosphatase, and tumor size greater than 3 cm results in higher relapse rate.	B	[28, 78-80]	III	retrospective
3.	**adjuvant radiotherapy when en bloc or total resection is not feasible or requires unacceptable functional sacrifices.** Radiotherapy seems to be an effective adjuvant treatment when total resection is not feasible	B	[78]	III	retrospective

Table 18. Solitary plasmocytoma

We recommend:

1.	**definite radiotherapy as a treatment of choice.** Stand-alone radiotherapy achieves very good results in terms of local control. Chemotherapy and/or novel therapies should be investigated for bone or bulky extramedullary tumors.	B	[81-83	III	retrospective

Table 19. Giant cell tumor.

We recommend:

1.	**surgical treatment (curettage or en bloc resection) as the mainstay of therapy.**	B	[84]	III	Case control study
2.	**Denosumab as a treatment of choice for the treatment of locally advanced tumors.**	B	[85-88]	III	Prospective comparative study
3.	**considering neoadjuvant therapy with Denosumab to achieve radical surgical treatment.**	B	[86-88]	III	Prospective comparative study

Table 20. Haemangioma with clinical manifestation

We recommend:

1.	**vertebroplasty for treatment of tumors with clinical manifestation.**	C	[89]	IV	Case series
2.	**considering radiotherapy is some cases.** Radiotherapy is safe, and effective in pain relief treatment for spinal haemangioma. Total doses of at least 34 Gy give the best symptomatic response.	B	[90]	III	Retrospective
3.	**radical surgical resection for hemangiomas with an extraosseous extension causing neurological symptoms.** Local recurrence of the tumor after subtotal resection has been reported, and adjuvant radiotherapy makes a second surgery difficult.	C	[91]	IV	Retrospective short case series

Table 21. Osteoid osteoma

We recommend:

1.	**conservative treatment**. Osteoid osteoma can be treated conservatively. Surgical excision (curettage) was considered the gold standard in the past and is no longer attractive today due to its invasiveness. Surgical resection should be taken into consideration as an option when the results of conservative treatment are poor.	A	[92]	I	Systematic review
2.	**Surgical resection, radiofrequency, percutaneous laser, and cryoablation in patients harboring painful spinal osteid osteoma or when the results of conservative treatment are poor**	B	[93-96]	III	Case series

Table 22. Osteochondroma

We recommend:

1.	**complete resection of the cartilaginous cap of the tumor.**	B	[97]	III	Case control, systematic review

Table 23. Aneurysmal bone cyst

We recommend:

1.	**selective arterial embolization as the first treatment option for spine aneurysmal bone cyst without neurologic deficit, pathological fracture or spinal instability.** It can be followed by surgery in case of reccurence/inefficiency	B	[98]	III	Retrospective study
2.	**complete intralesional excision as the therapy of choice in case of neurologic involvement, pathologic fracture, technical impossibility of performing embolization, or local recurrence after embolization procedures.**	B	[98, 99, 100, 101, 102]	III	Retrospective Systematic review

	Remark: radical surgical excision or en bloc resection are correlated with better prognosis for local tumor control with significantly lower recurrence rate especially when combined with the use of adjunctive therapies such as cryotherapy, phenol, or adjuvant radiotherapy. Recurrence rate ranked from the highest to the smallest in as follows: isolated surgiflo injection into the lesion, decompression/laminectomy, partial excision/resection, curettage alone. Primary or adjuvant radiotherapy may be an effective and safe treatment option for persistent or recurrent aneurysmal bone cyst				

Table 24. Giant cell tumor

We recommend:

1.	**complete surgical resection whenever possible particularly if neurologic impairment is present.** En bloc resection with wide/marginal margins should be performed when technically feasible because it is associated with decreased local reccurence. Intralesional resection is associated with increased local reccurence, and mortality correlates with local reccurence.	B	[103, 104]	III	ambispective observational study
2.	**Denosumab as neoadjuvant or adjuvant therapy where Enneking appropriate resection is not possible.** Denosumab alone is effective in relieving pain, increasing the ossification and sometimes reducing the tumor volume. It can be considered when surgical treatment cannot be radical due to associated unacceptable morbidity or loss of functions.	B	[105]	III	Prospective study

Table 25. Fibrous dysplasia

We recommend:

1.	**treatment with conventional surgical procedures including internal fixation.**	B	[106]	III	Literature review

Table 26. Langerhans cel histiocytosis

We recommend:

1.	**individual approach when establishing therapeutic management.** The spectrum of therapeutic possibilities is wide and, after the exclusion of a malignant lesion, it extends to non-surgical treatment, alternate administration of corticosteroids, curettage and replacement of the defect with bone grafts, and even surgical removal en block.	C	[107-109]	IV	Case series, review of case series