## Histological tumour grade (Core)

Histologic tumour grade offers important prognostic information. While several different grading systems exist, the French<sup>1</sup> grading system is the most widely used (see Table 2). This system is based on the assessment of differentiation, mitotic count, and necrosis.<sup>1</sup> Importantly, the system only applies to specific histotypes (see Table 3). Many other histotypes are not gradable. Reliable tumour grading is not possible after neoadjuvant therapy.

## Table 2: Tumour Differentiation Score According to Histologic Type in the Updated Version of the French Federation of Cancer Centers Sarcoma Group System.<sup>1</sup>

Histologic type	Score
Atypical lipomatous tumour/Well-differentiated liposarcoma	1
Well-differentiated leiomyosarcoma	1
Malignant neurofibroma	1
Well-differentiated fibrosarcoma	1
Myxoid liposarcoma	2
Conventional leiomyosarcoma	2
Conventional fibrosarcoma	2
Myxofibrosarcoma	2
High-grade myxoid (round cell) liposarcoma	3
Pleomorphic liposarcoma	3
Dedifferentiated liposarcoma	3
Pleomorphic rhabdomyosarcoma	3
Poorly differentiated/pleomorphic leiomyosarcoma	3
Biphasic/monophasic/poorly differentiated Synovial sarcoma	3
Mesenchymal chondrosarcoma	3
Extraskeletal osteosarcoma	3
Extraskeletal Ewing sarcoma	3
Malignant rhabdoid tumour	3
Undifferentiated pleomorphic sarcoma	3
Undifferentiated sarcoma, not otherwise specified	3

## Table 3: Guidelines for grading soft tissue sarcomas.

<ul> <li>Ewing sarcoma         <ul> <li>Rhabdomyosarcoma (all types)</li> <li>Angiosarcoma             <ul></ul></li></ul></li></ul>	Tumours which are by definition high grade	Tumours of varying behaviour for which	
<ul> <li>Angiosarcoma</li> <li>Pleomorphic liposarcoma</li> <li>Soft tissue osteosarcoma</li> <li>Mesenchymal chondrosarcoma</li> <li>Desmoplastic small cell tumour</li> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Ueiomyosarcoma</li> <li>Malignant peripheral nerve sheath tumour</li> <li>Solitary fibrous tumour</li> <li>Myxofibrosarcoma</li> <li>Dedifferentiated liposarcoma<sup>a</sup></li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Ewing sarcoma	grading or tumour-specific risk assessment	
<ul> <li>Pleomorphic liposarcoma</li> <li>Soft tissue osteosarcoma</li> <li>Mesenchymal chondrosarcoma</li> <li>Desmoplastic small cell tumour</li> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Well differentiated liposarcoma/atypical lipomatous tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Rhabdomyosarcoma (all types)	may be prognostically useful	
<ul> <li>Soft tissue osteosarcoma</li> <li>Mesenchymal chondrosarcoma</li> <li>Desmoplastic small cell tumour</li> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Solitary fibrous tumour</li> <li>Solitary fibrous tumour</li> <li>Solitary fibrous tumour</li> <li>Myxofibrosarcoma</li> <li>Dedifferentiated liposarcoma<sup>a</sup></li> <li>Tumours which are by definition low grade</li> <li>Well differentiated</li> <li>liposarcoma/atypical lipomatous</li> <li>tumour</li> <li>Epithelioid hemangioendothelioma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Etraskeletal myxoid chondrosarcoma</li> <li>Clear cell sarcoma</li> <li>Clear cell sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Angiosarcoma	Myxoid liposarcoma	
<ul> <li>Mesenchymal chondrosarcoma</li> <li>Desmoplastic small cell tumour</li> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Solitary fibrous tumour</li> <li>Myxofibrosarcoma</li> <li>Dedifferentiated liposarcoma/atypical lipomatous tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Pleomorphic liposarcoma	Leiomyosarcoma	
<ul> <li>Desmoplastic small cell tumour</li> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Solitary fibrous tumour</li> <li>Myxofibrosarcoma</li> <li>Dedifferentiated liposarcoma<sup>a</sup></li> <li>Tumours which are by definition low grade</li> <li>Well differentiated</li> <li>liposarcoma/atypical lipomatous</li> <li>tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma<sup>a</sup></li> <li>Yunours arcoma<sup>a</sup></li> </ul>	Soft tissue osteosarcoma	Malignant peripheral nerve sheath	
<ul> <li>Extra-renal rhabdoid tumour</li> <li>Intimal sarcoma</li> <li>Myxofibrosarcoma</li> <li>Dedifferentiated liposarcoma<sup>a</sup></li> <li>Tumours which are by definition low grade</li> <li>Well differentiated</li> <li>liposarcoma/atypical lipomatous</li> <li>tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Mesenchymal chondrosarcoma	tumour	
<ul> <li>Intimal sarcoma</li> <li>Intimal sarcoma</li> <li>Dedifferentiated liposarcoma<sup>a</sup></li> <li>Well differentiated liposarcoma/atypical lipomatous tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma<sup>a</sup></li> <li>Synovial sarcoma<sup>a</sup></li> </ul>	Desmoplastic small cell tumour	Solitary fibrous tumour	
Tumours which are by definition low grade       Tumours of varying behaviour for which         • Well differentiated       grading parameters are not yet well defined         • liposarcoma/atypical lipomatous       • Epithelioid hemangioendothelioma         • Dermatofibrosarcoma protuberans <sup>b</sup> • Extraskeletal myxoid chondrosarcoma         • Infantile fibrosarcoma       • Extraskeletal myxoid chondrosarcoma         • Alveolar soft part sarcoma       • Clear cell sarcoma         • Epithelioid sarcoma       • Synovial sarcoma <sup>a</sup> • 'Low-grade' fibromyxoid sarcoma       • Clear cell fibromyxoid sarcoma	Extra-renal rhabdoid tumour	Myxofibrosarcoma	
<ul> <li>Well differentiated liposarcoma/atypical lipomatous tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which ofter metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma<sup>a</sup></li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	Intimal sarcoma	Dedifferentiated liposarcoma <sup>a</sup>	
<ul> <li>liposarcoma/atypical lipomatous tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Mineratorial and the second second</li></ul>	Tumours which are by definition low grade	Tumours of varying behaviour for which	
<ul> <li>tumour</li> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Extraskeletal myxoid chondrosarcoma</li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	Well differentiated	grading parameters are not yet well defined	
<ul> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	liposarcoma/atypical lipomatous	Epithelioid hemangioendothelioma	
<ul> <li>Infantile fibrosarcoma</li> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	tumour	Extraskeletal myxoid chondrosarcoma	
<ul> <li>Tumours which are not gradable but which often metastasize within 10-20 years of follow-up</li> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	<ul> <li>Dermatofibrosarcoma protuberans<sup>b</sup></li> </ul>		
<ul> <li>Alveolar soft part sarcoma</li> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	Infantile fibrosarcoma		
<ul> <li>Clear cell sarcoma</li> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	Tumours which are not gradable but which often metastasize within 10-20 years of follow-up		
<ul> <li>Epithelioid sarcoma</li> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	<ul> <li>Alveolar soft part sarcoma</li> </ul>		
<ul> <li>Synovial sarcoma<sup>a</sup></li> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>	Clear cell sarcoma		
'Low-grade' fibromyxoid sarcoma	Epithelioid sarcoma		
	<ul> <li>Synovial sarcoma<sup>a</sup></li> </ul>		
Sclerosing onitholioid fibrosarsoma	<ul> <li>'Low-grade' fibromyxoid sarcoma</li> </ul>		
Scierosing epitheliolo incosarcoma	Sclerosing epithelioid fibrosarcoma		

<sup>a</sup> Some studies have shown prognostic difference between Grades 2 and 3 using the French grading system. <sup>b</sup> Fibrosarcomatous Dermatofibrosarcoma Protuberans (DFSP) is usually regarded as intermediate grade.

## Reference

1 Guillou L, Coindre JM, Bonichon F, Nguyen BB, Terrier P, Collin F, Vilain MO, Mandard AM, Le Doussal V, Leroux A, Jacquemier J, Duplay H, Sastre-Garau X and Costa J (1997). Comparative study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group grading systems in a population of 410 adult patients with soft tissue sarcoma. *J Clin Oncol* 15(1):350-362.