Table 1: Risk stratification for rhabdomyosarcoma

Risk group	**Sub-groups**	Pathology	Post surgical stage (IRS group)	Site	Node stage	Size & Age
Low	A	Favourable	I	Any	N0	Favourable
Standard	*B*	Favourable	I	Any	N0	Unfavourable
	C	Favourable	II, III	Favourable	N0	Any
	D	Favourable	II, III	Unfavourable	N0	Favourable
High	*E*	Favourable	II, III	Unfavourable	N0	Unfavourable
	F	Favourable	II, III	Any	N1	Any
	G	Unfavourable	I, II, III	Any	N0	Any
Very High	*H*	Unfavourable	II, III	Any	N1	Any

For sarcoma not otherwise specified (nos) see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.** and **Fehler! Verweisquelle konnte nicht gefunden werden.**.

For rhabdomyosarcoma in patients >21 years see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**.

Pathology: *Favourable* = All embryonal, spindle cells, botryoid RMS;

Unfavourable = All alveolar RMS (including the solid-alveolar variant).

Post surgical stage: (according to the IRS grouping system, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**):

Group I = Primary complete resection (R0);

Group II = Microscopic residuals (R1) or primary complete resection but N1;

Group III = Macroscopic residuals (R2).

Site: *Favourable* = Orbit, genito-urinary non bladder/prostate (i.e. paratesticular or vagina/uterus), non parameningeal head & neck;

Unfavourable = All other sites (parameningeal, extremities, genito-urinary bladder/prostate and "other site").

Node stage: (according to the TNM classification, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**):

N0 = No clinical or pathological node involvement;

N1 = Clinical or pathological nodal involvement.

Size & Age: *Favourable* = Tumour size (maximum dimension) ≤ 5 cm ***and*** age < 10 years;

Unfavourable = all others (i.e. size > 5 cm and / ***or*** age ≥ 10 years).

***Please note*:**

- Children with ascites/pleural effusion or CSF positive for malignant cells should be treated as stage IV patients (see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.** and below).
- For patients with synovial sarcoma, extraosseus Ewing's sarcoma, peripheral primitive neuroectodermal tumours or undifferentiated sarcoma, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.** and below.

Risk group	Pathology	Post surgical stage (IRS group)	Initial tumour size	Node stage
Localised RMS-like	SySa, STET (EES/pPNET), UDS	I, II, III	Any	Any
Metastatic disease		IV	Any	Any

Table 2: Risk stratification for rhabdomyosarcoma-like soft tissue sarcoma (SySa, STET, UDS)

Table 3: Risk stratification for "Non-RMS-like"-tumours

Risk Group	Histology	Node stage	IRS group	Initial tumour size
Low	Any (except MRT & DSRCT)*	N0	I	≤ 5 cm
Standard	Any (except MRT & DSRCT)*	N0	I	> 5 cm [1]
		N0	II	Any
		N0	III	≤ 5 cm [2]
High	MRT / DSRCT	N0 / N1	I, II, III	Any
	Any	N0	III	> 5 cm
	Any	N1	II, III	Any
Stage IV	Any	N0 / N1	IV	Any [3]

*** MRT** (malignant rhabdoid tumour), **DSRCT** (desmoplastic small and round cell tumour): treatment in the **High Risk Group**.
[1] Exception: Typical low grade tumours (grade 1) might be treated in the **Low Risk Group**.
[2] Exception: High grade tumours (grade 2 or 3) might be treated in the **High Risk Group**.
[3] Please refer to chapter **Fehler! Verweisquelle konnte nicht gefunden werden.** for treatment of stage IV (metastatic) soft tissue sarcoma.

- **Post surgical stage** (according to the IRS grouping system, chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Group I = Primary complete resection (R0), no microscopic tumour residuals;
  Group II = Microscopic tumour residuals (R1) or primary complete resection but N1;
  Group III = Macroscopic tumour residuals (R2).
- **Node stage** (according to the TNM classification, chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  N0 = No clinical or pathological node involvement;
  N1 = Clinical or pathological nodal involvement.
- **Initial tumour size** (according to the TNM classification, chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**):
  Favourable = Tumour size (maximum dimension) ≤ 5 cm (Ta);
  Unfavourable = Tumour size > 5 cm (Tb).

- **Pathology**
  Aggressive angiomyxoma (AAM)
  Angiomatoid fibrous histiocytoma (AFH)
  Alveolar soft part sarcoma (ASPS, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Chordoma (CHORD)
  Congenital (infantile) fibrosarcoma (cFS, please refer to chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Clear cell sarcoma (CCS)
  Dermatofibrosarcoma protuberans (DFSP, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Desmoplastic small and round cell tumour (DSRCT)
  Extraskeletal chondrosarcoma (ECS; including mesenchymal and myxoid CS, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)**
  Epithelioid sarcoma (ES)
  (Undifferentiated) Embryonal sarcoma of the liver (ESL) (should be treated as RME; for guidance refer to Study Centre)
  Endometrial stromal sarcoma (ESS)
  Fibrosarcoma (FS; see also below: congenital (infantile) fibrosarcoma)**
  Gastrointestinal stromal tumour (GIST, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Giant cell tumour, extraosseous (GCT)
  Inflammatory myofibroblastic tumour (IMT) and sarcoma (IMS, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Juvenile nasopharyngeal angiofibroma (JNF; see fibromatosis in chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Kaposi sarcoma (KS; please refer to the CWS Study Centre for an individual guidance)
  Low grade fibromyxoid sarcoma (LGFMS)
  Leiomyosarcoma (LMS)
  Lipoblastoma (LPB)
  Liposarcoma (LPS)
  Malignant ectomesenchymoma (MEM, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
  Malignant fibrous histiocytoma (MFH)
  Malignant mesenchymal tumour (MMM)
  Malignant peripheral nerve sheath tumour ((MPNST see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**),
  also neurofibrosarcoma (NFS) or malignant schwannoma)**

Malignant rhabdoid tumour (MRT)
Myofibroblastic sarcoma (MFS)
Myo-/Fibromatoses (see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
Myxoinflammatory fibroblastic sarcoma (MIFS)
Myxofibrosarcoma (MYX)
PEComa (PEC)
Pleuropulmonary blastoma (PPB, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)
Plexiform fibrohistiocytic tumour (PFT)
Pigmented neuroectodermal tumour of childhood (Retina Anlage Tumour, RAT)
Solitary fibrous tumour (SFT)
Undifferentiated pleomorphic high-grade sarcoma (UPS)
Vascular tumours (VS), such as hemangioendothelioma (HE), hemangiopericytoma (HP), angiosarcoma (AS, see chapter **Fehler! Verweisquelle konnte nicht gefunden werden.**)