Surgical Approach to Non-Rhabdomyosarcoma Soft Tissue Sarcoma in Children

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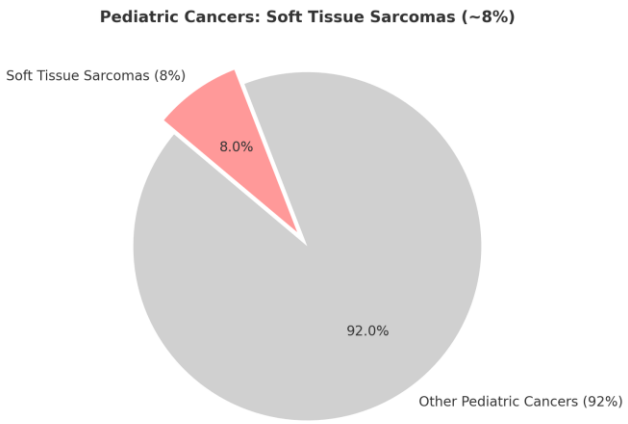
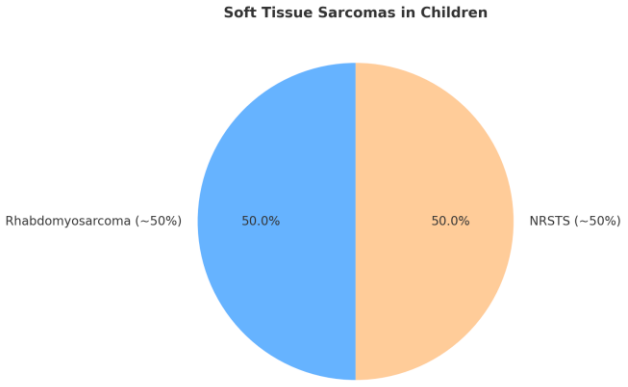
Epidemiology & Background



- Soft tissue sarcomas = 8% of pediatric cancers
- ≈50% are NRSTS, >50 histologic subtypes
- Prognosis depends on size, grade, site, margins, metastasis
- Less chemosensitive → surgery central



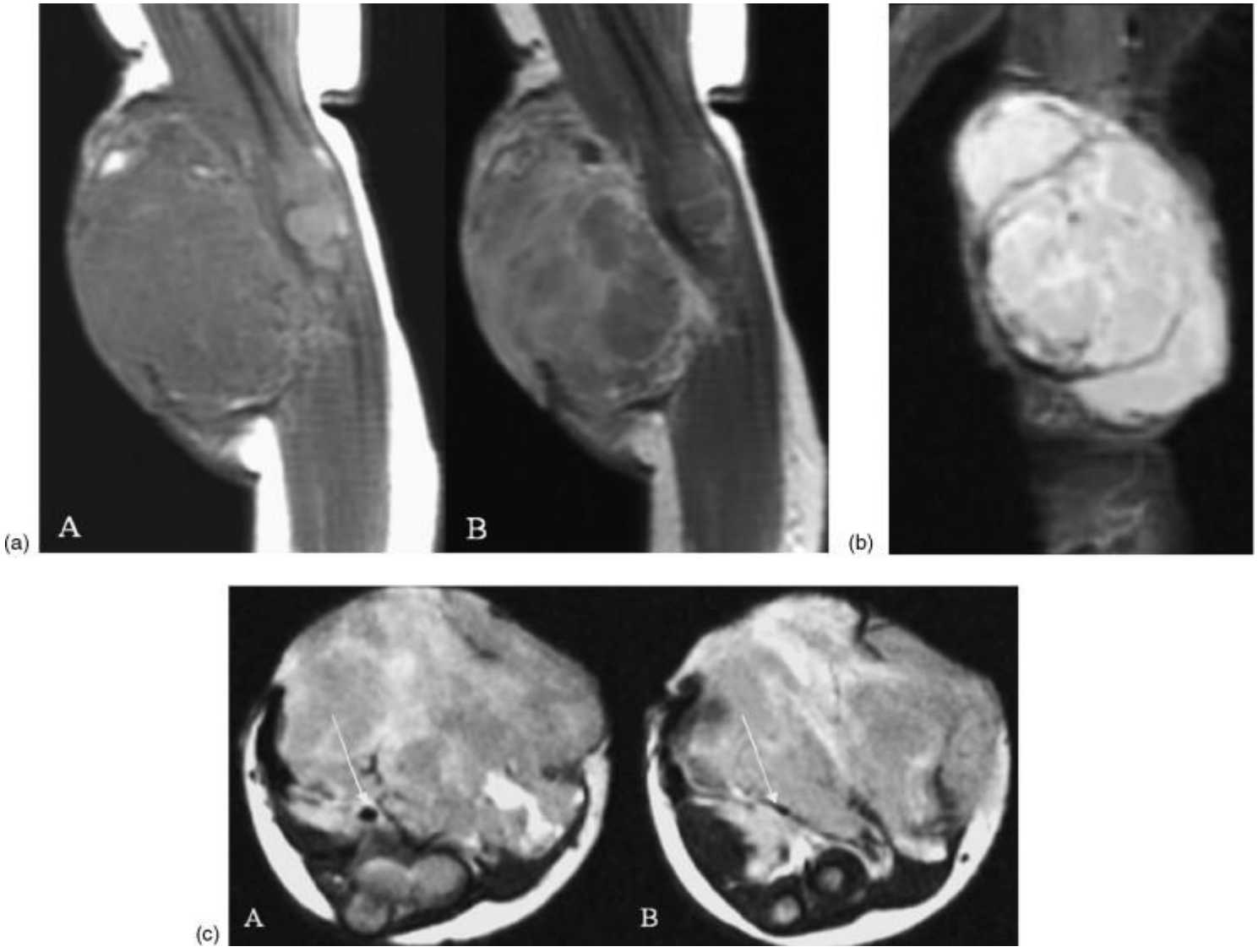
Prevalent Pediatric NRSTS

- **Infantile Fibrosarcoma:** ETV6–NTRK3 fusion, good prognosis in infants
- **Synovial Sarcoma** : SS18–SSX fusion, outcome by size/invasion
- **MPNST** : NF1 association, surgery crucial
- **DSRCT** : peritoneal disease, cytoreduction ± HIPEC
- **Desmoid Tumor** : aggressive but non-metastatic, observe vs surgery
- **DFSP** : COL1A1–PDGFB fusion, wide excision, imatinib option

Work-up & Multidisciplinary Planning

- MRI of primary site (local extent, NV involvement)
- CT chest for lung metastases
- Core-needle biopsy (tract resected en bloc)
- Multidisciplinary tumor board before surgery

Infantile Fibrosarcoma



Synovial Sarcoma

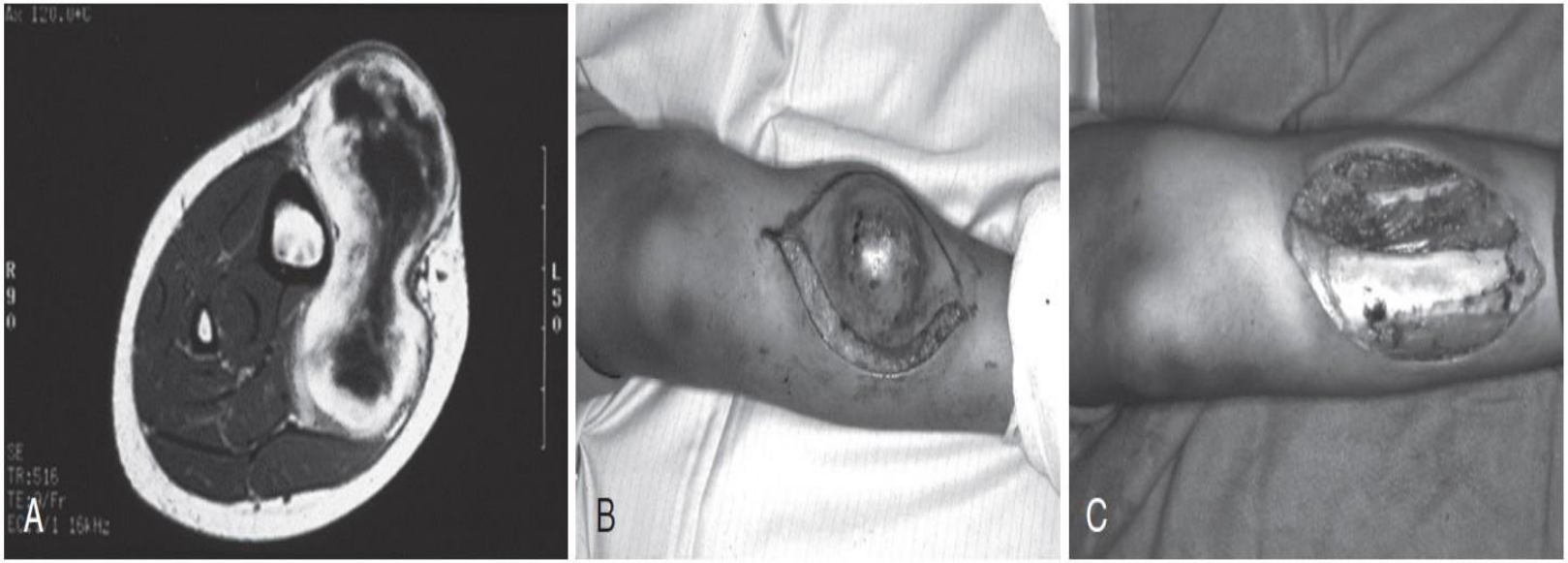


FIGURE 36-1 **A-C**, Magnetic resonance (MR) image of a child with synovial sarcoma abutting the tibia. Neoadjuvant chemotherapy was not successful in reducing the size of the tumor. Marginal resection with postoperative radiation or brachytherapy is a preferred alternative to amputation.

MPNST

DFSP

Risk Stratification (COG ARST0332)

- **Low risk**: R0/R1 low-grade; small high-grade
- **Intermediate**: large/high-grade; R1/R2; unresected non-metastatic
- **High risk**: metastatic disease
- Guides needed for surgery ± RT ± chemo

Surgical Margin Principles

- **Goal**: R0 resection, preserve function
- **INSTRuCT**: R0 / R1 / R2 + margin distance
- Planned close margins acceptable near critical structures
- No fixed width requirement; tailor to case

When to Add Radiation

- R1 resections in high-grade or >5 cm tumors
- Pre-op RT: improves resectability, spares growth plates
- Post-op RT: for unexpected positive margins
- Modalities: IMRT, proton, brachytherapy

Role of Chemotherapy & Targeted Therapy

- Limited role overall
- Neo-adjuvant chemo for unresectable/large tumors
- Histology-specific: e.g. synovial sarcoma, DSRCT
- Targeted: TRK inhibitors (IFS), imatinib (DFSP)

Special Surgical Scenarios

- Pulmonary metastases → resection (thoracotomy/VATS)
- Sentinel node biopsy: epithelioid, clear cell, some SS
- Local recurrence → re-excision + RT if not irradiated
- Amputation = last resort

Prognostic Factors

- Tumor ≥5 cm
- High grade
- Intra-abdominal location
- Positive margins
- Age: infants best, adolescents worse

Histology-Specific Pearls

- IFS: surgery ± TRK inhibitors
- SS: R0 resection, RT for margins, chemo selective
- MPNST: radical resection, poor chemo effect
- DSRCT: cytoreduction + HIPEC
- Desmoid: observe first, operate only if progression
- DFSP: wide excision or Mohs, imatinib if unresectable

Hyperthermic Intraperitoneal Chemotherapy (HIPEC)

- Used mainly in Desmoplastic Small Round Cell Tumor (DSRCT)
- Combines cytoreductive surgery with heated chemotherapy perfusion
- Targets microscopic peritoneal disease left after debulking
- Early pediatric experience: ↑ disease-free survival, though morbidity exists
- Currently performed only in specialized centers

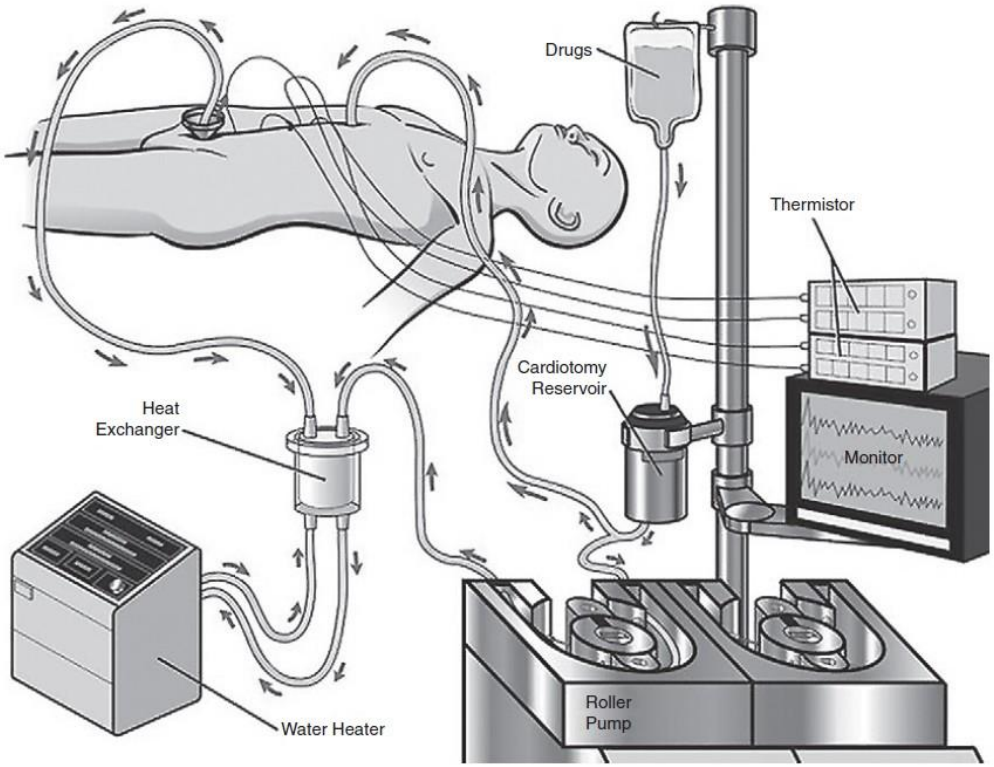


FIGURE 36-3 Setup for hyperthermic intraperitoneal chemotherapy (HIPEC) therapy for children with "sarcomatosis" after cytoreductive surgery.

Key Take-Home Messages

- **Surgery** = cornerstone of cure
- **Aim:** R0 resection, preserve function
- **RT** for positive/close margins and large high-grade tumors
- **Chemo/targeted** = selective
- **MDT** care + trial enrollment improve outcomes

References

- Coran’s Pediatric Surgery, 7th ed.
- COG ARST0332 trial
- EpSSG NRSTS guidelines
- INSTRuCT surgical margin consensus
- NCI PDQ Pediatric NRSTS (2025 update)

Thank You !