Management of Pediatric Fibrosarcoma

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Overview

- Rare soft tissue sarcoma in children
- Two forms: Infantile (congenital) vs. Older children (adult-type)
- Distinct biology → distinct management

Infantile Fibrosarcoma (IFS)

- Most common soft tissue sarcoma <1 yr
- Rapidly growing but rarely metastasizes
- Pathognomonic ETV6–NTRK3 fusion

ETV6 --- NTRK3
(t12;15) Fusion

IFS – Standard Management

- Surgery: cornerstone, aim for limb-sparing resection
- Chemotherapy: VA regimen (vincristine, actinomycin D) highly effective
- Radiation: rarely needed

IFS – Treatment Strategy

- Primary complete resection → observation
- Unresectable/large tumor → neoadjuvant chemo or TRK inhibitors
- R1 resection: close follow-up; some regress spontaneously

IFS Treatment Strategy:

Resectable → Surgery → Observation
Unresectable → Chemo / TRK Inhibitors → Surgery
R1 Resection → Observation / Chemo

IFS – Targeted Therapy

- Larotrectinib / Entrectinib
- ORR >90%, rapid responses, limb-sparing effect
- Changing frontline therapy paradigm

Before
(TRK+ tumor)

After
(Larotrectinib)

Fibrosarcoma in Older Children

- Rare, biologically aggressive
- No NTRK fusion
- Higher risk of metastasis (lungs)

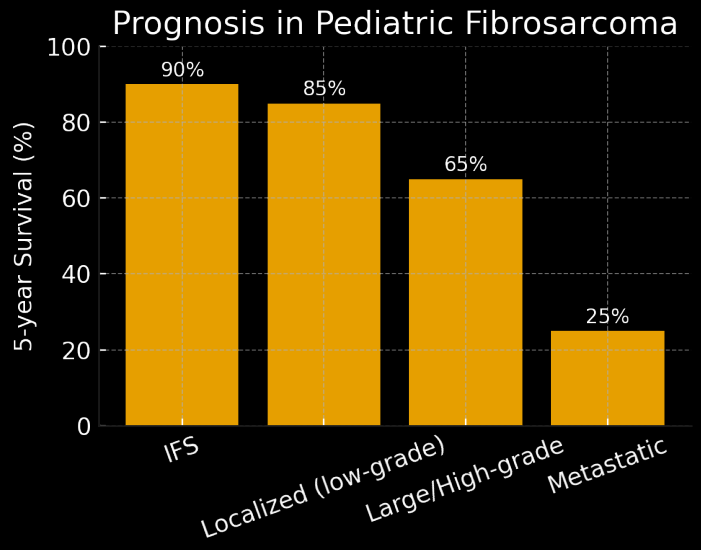
Surgery (wide margins)
├─ Negative margins → Observe / ±RT
└─ Positive margins → RT ± Chemo
High-grade / Large / Metastatic → Ifosfamide + Doxorubicin

Older Children – Standard Management

- Surgery: wide margins, limb-sparing if possible
- Radiation: for R1 or unresectable tumors
- Chemotherapy: ifosfamide + doxorubicin (neoadjuvant/ adjuvant)

Emerging Therapies

- COG ARST1321: pazopanib + chemoradiation → ↑ necrosis
- EpSSG upcoming trial: regorafenib + ifosfamide/doxorubicin
- Immunotherapy: still under investigation



Prognosis

- Infantile: excellent (5-yr survival 85–100%)
- Older children: Localized low-grade: 80–90%
- Large/high-grade: 60–80%, Metastatic: 20–35%

0-2 yrs

Every 3-4 months

Follow-Up

- Chest imaging every 3–4 mo (first 2 yrs)
- Primary site MRI/US as indicated
- Survivorship: growth, cardiac, fertility monitoring

Take-Home Messages

- Infantile: chemosensitive, NTRK-targetable, excellent survival
- Older children: aggressive, multimodal therapy needed
- TRK inhibitors = paradigm shift in IFS
- International collaboration vital for rare tumors

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