

Disclaimer: This algorithm has been developed for MD Anderson using a multidisciplinary approach considering circumstances particular to MD Anderson's specific patient population, services and structure, and clinical information. This is not intended to replace the independent medical or professional judgment of physicians or other health care providers in the context of individual clinical circumstances to determine a patient's care. This algorithm should not be used to treat pregnant women.

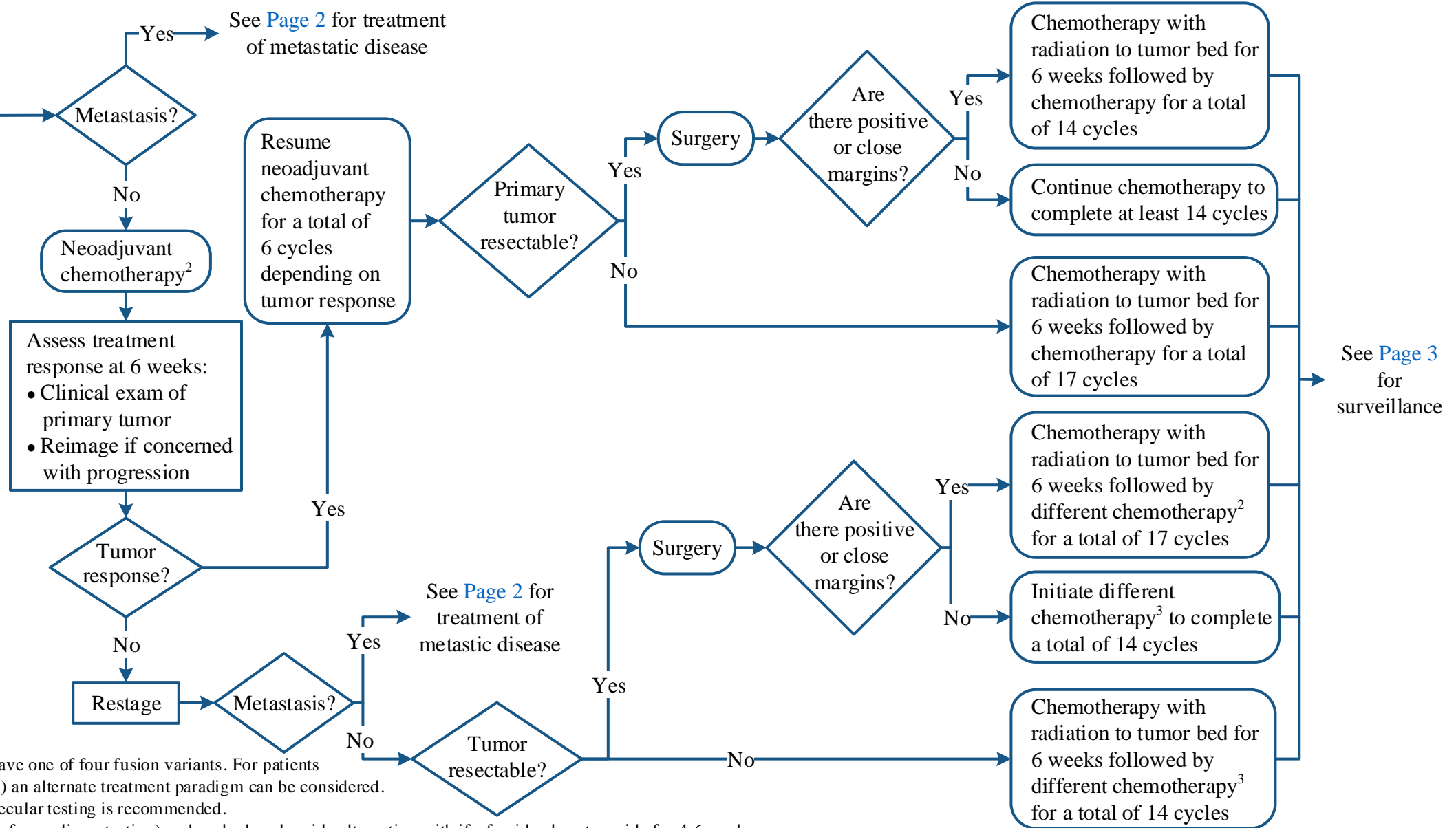
Note: Consider Clinical Trials as treatment options for eligible patients. Referral to a center with both pediatric oncology and orthopedic surgery is essential.

CLINICAL EVALUATION

- History and physical
- CBC with differential, total protein, albumin, calcium, total bilirubin, alkaline phosphatase, LDH, AST, sodium, potassium, chloride, carbon dioxide, and PT/INR
- Plain films of primary tumor
- MRI of primary tumor
- Bone scan
- Chest x-ray
- CT chest
- Whole body PET/CT (optional)
- Biopsy (open vs. core needle)
- Histology review by bone tumor pathologist
- Screening MRI of spine or bone marrow biopsy and aspirate
- EKG and cardiac scan (ECHO)
- Central venous access device (CVAD) placement
- Pregnancy test if clinically indicated
- Discuss fertility
- Cytogenetics and/or molecular studies (may require re-biopsy)¹

PRIMARY TREATMENT

ADJUVANT TREATMENT



¹ Greater than 95% of Ewing sarcoma will have one of four fusion variants. For patients with Ewing-like sarcoma (e.g., CIC-DUX4) an alternate treatment paradigm can be considered. For those who are negative, additional molecular testing is recommended.

² Vincristine, doxorubicin (with dexrazoxane for cardioprotection) and cyclophosphamide alternating with ifosfamide plus etoposide for 4-6 weeks

³ Temozolomide plus irinotecan (5 days every 3 weeks), or clinical trial if available

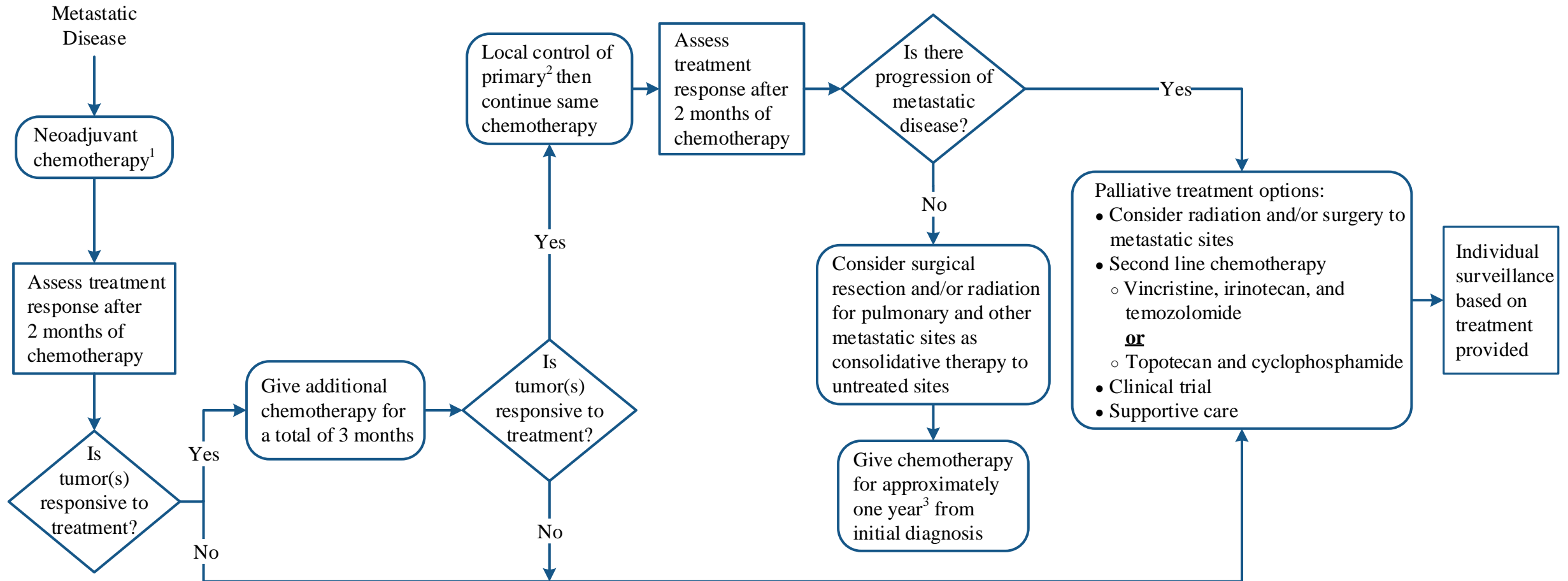
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PRIMARY TREATMENT

ADJUVANT TREATMENT

SURVEILLANCE



¹ Vincristine, doxorubicin (with dexrazoxane for cardioprotection) and cyclophosphamide alternating with ifosfamide plus etoposide for 4-6 weeks

² Local control: axial lesions undergo radiation, extremity lesions undergo surgery and/or radiation, and head and neck lesions are treated individually based on clinical indications

³ Monitor for progression after 2-3 months of chemotherapy for approximately 1 year of treatment. If no progression of disease following completion of chemotherapy regimen then move patient to surveillance (see [Page 3](#)).

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Pediatric Ewing's Family of Tumors Surveillance

Total years for Surveillance				Year 1				Year 2			Year 3			Year 4		Year 5
Frequency of Surveillance by month	3	6	9	12	15	18	21	24	28	32	36	40	44	48	54	60
History and physical	x	x	x	x	x	x	x	x	x	x	x	x	x	x		x
Monitor and discuss with patient late effects of primary treatment	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x
CBC with differential		x		x		x		x		x		x		x		x
Total protein, albumin, calcium, phosphate, magnesium, glucose, creatinine, total bilirubin, alkaline phosphatase, LDH		x		x		x		x		x				x		x
Plain films of primary	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x
Pelvic primaries: MRI, pelvic x-ray	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x	x
Bone scan and/or PET scan for symptomatic patients with and/or without history of bone metastases	x	x	x	x		x		x			x					
Chest x-ray	x	x	x	x	x	x	x	x	x	x	x		x	x	x	x
CT chest (higher risk patients)	x	x	x	x				x			x			x		x

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SUGGESTED READINGS

Anderson, P., Kopp, L., Anderson, N., Cornelius, K., Herzog, C., Hughes, D., & Huh, W. (2008). Novel bone cancer drugs: Investigational agents and control paradigms for primary bone sarcomas (Ewing's sarcoma and osteosarcoma). *Expert Opinion on Investigational Drugs*, 17(11), 1703-1715. <https://doi.org/10.1517/13543784.17.11.1703>

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DEVELOPMENT CREDITS

This practice algorithm is based on majority expert opinion of the Pediatric Center Faculty at the University of Texas MD Anderson Cancer Center. It was developed using a multidisciplinary approach that included input from the following:

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