Single-system disease

- Bone-only –Single bone – Curettage provides tissue diagnosis and treatment. Radiation therapy (RT) may be used for selected adults, but not children.
- Multiple bones – For ≥2 bone lesions, lesion ≥5 cm, femoral or vertebral involvement, or CNS-risk bone (ie, orbit, mastoid, temporal, sphenoid), treatment involves systemic therapy.

Surgery or RT may be added in selected cases.

- Skin-only – Topical steroids or mustard, or oral hydroxyurea, methotrexate, thalidomide, or lenalidomide can be effective.
- Multisystem – Multisystem disease requires systemic therapy.
- Children – For initial systemic treatment of children with LCH, we suggest induction therapy with vinblastine plus prednisone (V-P), rather than other chemotherapy regimens or a targeted agent (Grade 2C).

Treatment response guides further management; continuation therapy is 12 months for response to V-P.

- CNS or risk organ involvement – For adults with BRAF V600E-mutated LCH and involvement of CNS or a risk organ, we suggest a BRAF inhibitor (eg, vemurafenib, dabrafenib), rather than systemic chemotherapy (Grade 2C).

For adults with BRAF wildtype LCH with CNS or risk organ involvement, we suggest cytarabine or cladribine, rather than combination chemotherapy or a targeted agent (Grade 2C).

- Response assessment – Positron emission tomography (PET) is preferred for response assessment, but computed tomography (CT), magnetic resonance imaging (MRI), or clinical assessment is used when PET is not available or appropriate (eg, brain lesions).
- **Long-term surveillance** – Patients are at risk for treatment-related toxicity, second cancers, and endocrine complications.

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Sp04

THROMBOSIS IN CHILDHOOD LEUKEMIA AND LYMPHOMA

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Thrombosis in childhood usually develops secondary to underlying causes. One of the most important risk factors is cancer. It has been reported that the incidence of thrombosis in children with cancer is 2-16% when symptomatic thrombosis is mentioned and it climbs up to 50% if asymptomatic conditions are included. Thrombosis associated with childhood cancers is multifactorial. In addition to the prothrombotic effect of cancer, mass effect, vascular invasion of cancer, drugs used (e.g., steroid, asparaginase), catheter, infection, immobilization, surgery, total parenteral nutrition, and comorbid genetic thrombophilia are the most important underlying etiologies. Thrombosis can cause morbidity, mortality, as well as inadequate or delayed treatment. Among childhood cancers, thrombosis risk is more common in acute lymphoblastic leukemia and lymphoma than in solid malignancies. Among the drugs used for the treatment of thrombosis, low molecular weight heparin constitutes the most important group. Warfarin, on the other hand, can be preferred in case of long-term use, but its use may be challenging due to polypharmacy and nutritional instability on warfarin efficiency. Thrombolytic therapies are rarely used in selected cases. In addition to general measures to reduce the risk of thrombosis, prophylaxis is controversial. Prophylaxis has not been included in the standard guidelines for the prevention of thromboembolic complications in childhood. It can be considered for use in high-risk patients. However, prophylaxis during cancer treatment may be more challenging, especially in this group of patients who need frequent interventions (e.g., intrathecal treatments) and have an increased risk of bleeding secondary to thrombocytopenia and coagulopathies. There are many continuing studies on the prophylactic and therapeutic use of new-generation anticoagulants in childhood.

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Sp05

CANCER TREATMENT-RELATED CARDIOTOXICITY: OPTIMIZING HEART HEALTH FROM DIAGNOSIS TO LONG-TERM FOLLOW-UP

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Improvements in overall survival rates for children diagnosed with cancer have led to a growing number of long-term childhood cancer survivors and an increasing recognition of the late health conditions they may experience. Among these are cardiac conditions, most commonly associated with prior anthracycline chemotherapy and chest-directed radiation exposing the heart. Potential late effects of anthracycline chemotherapy and chest-directed radiation therapy include cardiomyopathy, subclinical left ventricular dysfunction, heart failure, and arrhythmia. In addition, chest-directed radiation exposing cardiac substructures has been associated with risk for pericarditis, pericardial fibrosis, valvular disease, atherosclerotic heart disease and myocardial dysfunction. Patient (e.g., age at exposure, family history, genetic variation) and treatment (e.g., cumulative dose, multimodality cardiotoxic therapy) factors influence the magnitude of risk. In addition, co-morbid medical conditions (e.g., hypertension, diabetes, dyslipidemia, obesity) and health behaviors (e.g., smoking) can exacerbate risk in aging survivors. Recognition of treatment associations and adverse cardiac outcomes has informed risk-stratification strategies used in contemporary protocols and guided health surveillance recommendations for long-term survivors. Dexrazoxane has also been used for primary prevention of anthracycline cardiotoxicity in high