Burkitt and Double-Hit or Triple-Hit Lymphomas

Note: Consider Clinical Trials as treatment options for eligible patients.

DIAGNOSIS

ESSENTIAL:

- Hematopathology review of all slides with at least one paraffin block representative of the tumor. Rebiopsy if consult material is non-diagnostic. FNAs are generally inadequate. Recommend core or excisional biopsy.
- Adequate immunophenotyping to establish diagnosis
  - Paraffin Panel: CD3, CD10, CD20, CD45 (LCA), Ki-67, BCL2, BCL6, TdT
  - Flow cytometry immunophenotyping (optional if paraffin IHC has been performed): kappa/lambda light chains, IgM, CD3, CD5, CD10, CD19, CD20, CD45, TdT
  - In situ hybridization: EBER
- Molecular genetic analysis
  - For Burkitt lymphoma: Conventional cytogenetics helpful if available; FISH to detect *MYC* gene rearrangements
  - For Double-hit or Triple-hit lymphoma: FISH to detect *MYC* gene rearrangements. If positive, then check BCL2 and BCL6 gene rearrangements

STRONGLY RECOMMENDED:

- FNA or core biopsy for tissue banking by protocol
- Perform gene mutation panel if available

INITIAL EVALUATION

ESSENTIAL:

- Physical exam
- Performance status (ECOG)
  - B symptoms (Unexplained fever >38°C during the previous month; Recurrent drenching night sweats during the previous month; Weight loss >10 percent of body weight ≤ 6 months of diagnosis)
- CBC with differential, albumin, AST, ALT, total bilirubin, alkaline phosphorus, serum calcium, uric acid, phosphate, magnesium, BUN, creatinine, LDH
- Screening for HIV-1 and HIV-2, hepatitis B and C (HBcAb, HBsAg, HCV Ab)
- Chest X-ray, PA and lateral
- CT with contrast of neck, chest, abdomen and pelvis
- Echo or MUGA
- Lumbar puncture with cytology evaluation
- Bilateral bone marrow biopsy with aspirate
- PET/CT Scan
- Lifestyle risk assessment[1]

OF USE IN SELECTED CASES:

- Upper GI/barium enema/endoscopy
- MRI of brain with gadolinium or CT of brain
- Pregnancy test in women of childbearing potential
- Discussion of fertility issues and sperm banking

See Page 2, Clinical Presentation and Primary Treatment

[1] See Physical Activity, Nutrition, and Tobacco Cessation algorithms; ongoing reassessment of lifestyle risks should be a part of routine clinical practice

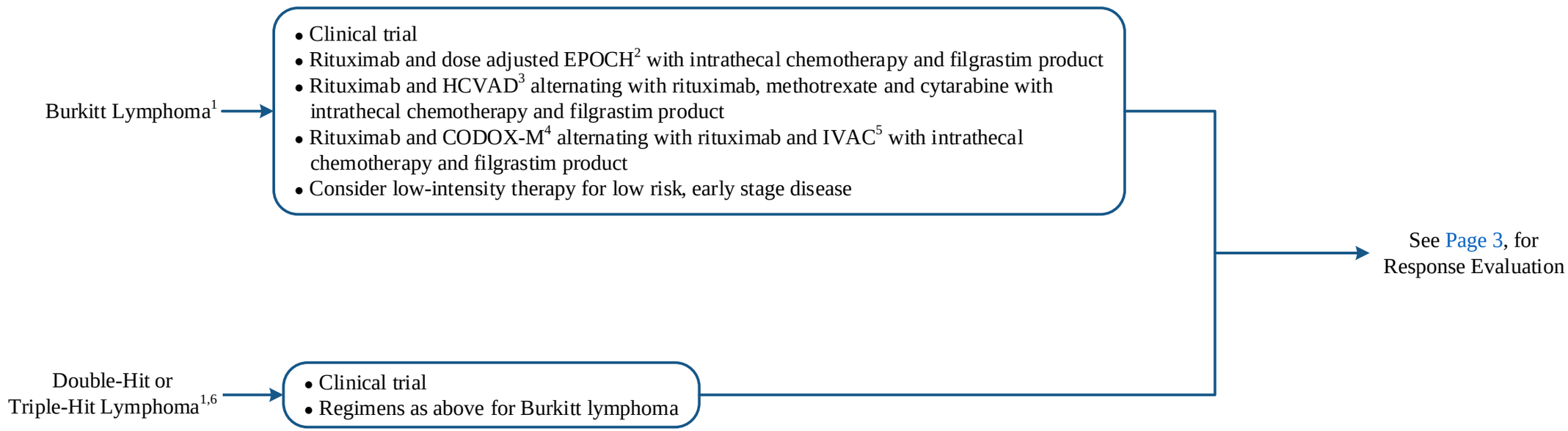
Department of Clinical Effectiveness V5
Approved by the Executive Committee of the Medical Staff on 10/15/2019

Burkitt and Double-Hit or Triple-Hit Lymphomas

Note: Consider Clinical Trials as treatment options for eligible patients.

CLINICAL PRESENTATION

PRIMARY TREATMENT



[1] CHOP: cyclophosphamide, doxorubicin, vincristine, and prednisone is not adequate therapy
[2] EPOCH: etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin
[3] HCVAD: cyclophosphamide, vincristine, doxorubicin, and dexamethasone
[4] CODOX-M: cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate and leucovorin
[5] IVAC: ifosfamide, etoposide, and high-dose cytarabine
[6] Also known as high grade B-cell lymphoma with *MYC* and/or BCL2 or BCL6 gene erarrangements

Burkitt and Double-Hit or Triple-Hit Lymphomas

Note: Consider Clinical Trials as treatment options for eligible patients.

RESPONSE EVALUATION

Complete response (CR) →

Recommend to continue:
- Routine cancer screening tests with Primary Cancer physician
- Year 1: every 3-4 months
  - Physical exam and labs
  - Repeat CT with contrast
- Year 2: every 6 months
  - Physical exam and labs
  - Repeat CT with contrast
- Years 3-5: every 12 months
  - Physical exam and labs
  - Repeat CT with contrast
- Year 5 and beyond: every 12 months
  - Physical exam and labs

Partial response (PR), stable disease, progressive disease and recurrence →

- Clinical trial
- Consider non-overlapping chemotherapy option per Diffuse Large B-Cell Lymphoma guidelines
- Consider high dose chemotherapy plus autologous stem cell transplant for patients who enter into second remission with good performance status and well controlled concomitant medical issues

SUGGESTED READINGS

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DEVELOPMENT CREDITS

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