Oncology Clinical Pathways
Primary Mediastinal B-Cell Lymphoma (PMBCL)

September 2024 – V2.2024

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Primary Mediastinal B-Cell Lymphoma – Presumptive Conditions

VA automatically presumes that certain disabilities were caused by military service. This is because of the unique circumstances of a specific Veteran's military service. If a presumed condition is diagnosed in a Veteran within a certain group, they can be awarded disability compensation.

Atomic Veterans – Exposure to Ionizing Radiation

- Lymphomas, other than Hodgkin's disease

Vietnam Veterans – Agent Orange Exposure or Specified Locations

- Non-Hodgkin's lymphoma

Gulf War and Post 9/11 Veterans

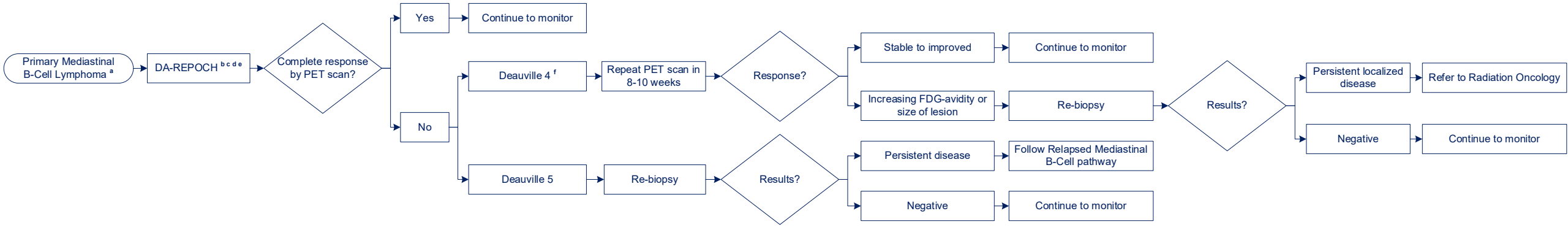
If the patient served on or after Sept. 11, 2001, in Afghanistan, Djibouti, Egypt, Jordan, Lebanon, Syria, Uzbekistan, or Yemen or if the patient served in the *Southwest Asia theater of operations, or Somalia, on or after Aug. 2, 1990, specific conditions include:

- Lymphoma of any type

* The Southwest Asia theater of operations refers to Iraq, Kuwait, Saudi Arabia, the neutral zone between Iraq and Saudi Arabia, Bahrain, Qatar, the United Arab Emirates, Oman, the Gulf of Aden, the Gulf of Oman, the Persian Gulf, the Arabian Sea, the Red Sea, and the airspace above these locations.

For more information, please visit U.S. Department of Veterans Affairs - Presumptive Disability Benefits (va.gov)

Primary Mediastinal B-Cell Lymphoma



Clinical trial(s) always considered on pathway. For assistance finding a clinical trial, email CancerClinicalTrialsNavigation@va.gov.

[a] **Initial Diagnosis** confirmation by biopsy should be excisional; core needle biopsy may be sufficient depending on the sample obtained; confirm pathology evaluation included Cell of Origin testing (CD10, MUM1, and BCL6 IHC) and evaluation for double hit +/- double expressor status (MYC, BCL2, and BCL6 break apart FISH; MYC and BCL2 IHC); consider additional pathology evaluation including cyclin D1, Ki-67, PAX5, CD30, CD15, MAL, CD200, PDL1, PDL2, EBER based on clinical situation after discussion with your pathologist; typical morphology shows large cells in clusters or sheets, frequently with a background of delicate fibrosis; PMBCL can be morphologically similar to DLBCL, cHL, and grey zone lymphoma; second review by a hematopathologist may be helpful; presence of non-mediastinal or bone marrow disease is usually not consistent; classification as PMBCL is based on clinical presentation; typical presentation is with an isolated anterior mediastinal mass; bone marrow biopsy is helpful to rule out involvement, which is rare in PMBCL

[b] **DA-REPOCH** recommend 6 cycles; labs (CBC/diff) should be checked 2x/week after each cycle for appropriate dose adjustments based on toxicity; refer to published literature for details; RCHOP +/- radiation is a reasonable alternative for those who may not tolerate REPOCH (e.g., age > 60, slightly frail, comorbidities); in situations where RCHOP alone is used, consolidation radiation maybe necessary to get to a complete remission and a post treatment PET scan is required to guide this decision

[c] **Blood Tests** include CBC, CMP, LDH, uric acid, Phos, Hep C Ab, Hep B sAg, Hep B sAb, Hep B cAb, HIV

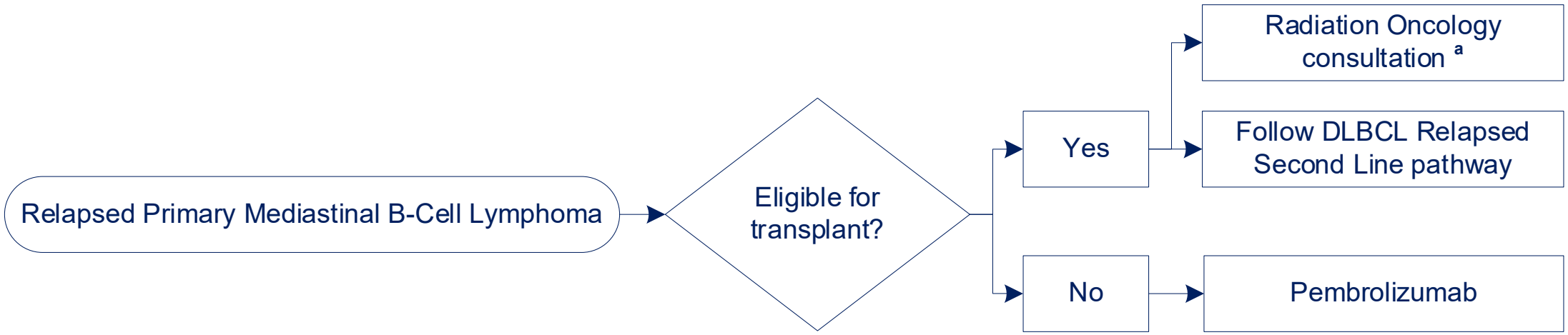
[d] **Cardiac Function** should be evaluated by echocardiogram or MUGA; good cardiac function defined as EF >50%; poor cardiac function defined as EF ≤ 50%

[e] **Supportive Care** empiric GCSF support should be used if age >65 years, cytopenias at diagnosis, bone marrow involvement; GCSF should be added if not already used if infections or febrile neutropenia occurs during therapy; anti-infection prophylaxis: VZV/HSV recommended; stimulant laxatives and anti-emetics recommended; consider inpatient monitoring and management for tumor lysis syndrome at cycle 1 in patients with high burden of disease, renal dysfunction, rapidly growing lymphoma; use allopurinol, intravenous fluids, and rasburicase as needed; consider inpatient monitoring for patients with intestinal involvement in cycle 1 due to risk of perforation; consider referral for fertility preservation for appropriate and interested patients; immunizations with pneumococcal and COVID vaccines recommended after chemotherapy; referral to Registered Dietitian for medical nutrition therapy

[f] **Deauville 4 Responses** with FDG uptake minimally above liver is common in PMBCL; a Deauville 4 response with FDG uptake markedly above liver may warrant an immediate biopsy

cHL Classic Hodgkin Lymphoma
DA R-EPOCH Dose-Adjusted Rituximab, Etoposide, Prednisone, Vincristine, Cyclophosphamide, Doxorubicin

Relapsed Primary Mediastinal B-Cell Lymphoma



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[a] **Radiation Oncology Consultation** to discuss post transplant radiation therapy

DLBCL Diffuse Large B-Cell Lymphoma

Primary Mediastinal B-Cell Lymphoma – Molecular Testing Table

Eligibility	Test Category	Test Type	Recommended Vendors	NPOP Coverage	Specimen Type
Primary Mediastinal Large B-Cell Lymphoma, Initial Diagnosis	IHC	Hans algorithm testing for cell of origin CD10, MUM1, and BCL6	Local VA or locally contracted vendor	No	Tissue (FFPE)
	IHC	Consider IHC for CD30 (if considering brentuximab therapy)	Local VA or locally contracted vendor	No	Tissue (FFPE)
	ISH	EBER in-situ hybridization	Local VA or locally contracted vendor	No	Tissue (FFPE)
	FISH	MYC, BCL2, BCL6 break apart FISH to exclude high grade B-cell lymphoma	Local VA or locally contracted vendor	No	Tissue (FFPE)