MAYO CLINIC LABORATORIES

Test Definition: SZDIA

Sezary Diagnostic Flow Cytometry, Blood

Overview

Useful For

Identifying immunophenotypically aberrant T-cell populations with restricted expression of T-cell receptor beta-chain constant in peripheral blood, to roughly assess the circulating tumor burden in cutaneous T-cell lymphomas

Reflex Tests

Test Id	Reporting Name	Available Separately	Always Performed
FCIMS	Flow Cytometry Interp, 9-15 Markers	No	No
FCINS	Flow Cytometry Interp,16 or greater	No	No

Additional Tests

Test Id	Reporting Name	Available Separately	Always Performed
FIRST	Flow Cytometry, Cell Surface, First	No	Yes
ADD1	Flow Cytometry, Cell Surface, Addl	No	Yes

Testing Algorithm

This Sezary panel is ordered for patients with a clinical suspicion of Sezary syndrome or cutaneous T-cell lymphoma with peripheral blood involvement **without** a previously confirmed diagnosis. A Triage panel and Sezary panel will always be performed. This test is **not indicated** for monitoring peripheral blood involvement in patients with a diagnosis of Sezary syndrome or mycosis fungoides. For monitoring purposes, order SZMON / Sezary Monitoring Flow Cytometry, Blood.

The panel is charged based on number of markers tested (FIRST for first marker, ADD1 for each additional marker). In addition, reflex testing may occur to fully characterize a disease state or clarify any abnormalities from the screening test. Reflex tests will be performed at an additional charge for each marker tested (ADD1 if applicable).

The testing process begins with a triage panel to evaluate for and exclude monotypic B cells or increased numbers of blasts. The triage panel also includes antibodies to assess the number of CD3-positive T cells and CD16-positive/CD3-negative natural killer cells present. Additional reflex testing may occur to fully characterize a disease state per algorithmic approach or clarify any abnormalities from the screening test at an additional charge for each marker tested (ADD1 if applicable). For a comprehensive list of potential additional panels, refer to LCMS Leukemia/Lymphoma Immunophenotyping, Flow Cytometry, Varies.

The Sezary panel will further evaluate the T cells for expression of CD2, CD4, CD5, CD7, CD8, CD26, and TRBC1.

Method Name

Immunophenotyping

NY State Available

Yes

Specimen

Specimen Type

Whole blood

Ordering Guidance

This test is **not indicated** for monitoring patients with a diagnosis of Sezary syndrome. For monitoring purposes, order SZMON / Sezary Monitoring Flow Cytometry, Blood.

Specimen Required

Container/Tube:

Preferred: Yellow top (ACD solution A or B)

Acceptable: Lavender top (EDTA), green top (sodium heparin)

Specimen Volume: 6 mL

Collection Instructions:

1. Send whole blood specimen in original tube. **Do not aliquot**.
2. Label specimen as blood.

Forms

If not ordering electronically, complete, print, and send a Hematopathology/Cytogenetics Test Request (T726) with the specimen.

Specimen Minimum Volume

1 mL

Reject Due To

Gross hemolysis	Reject
Gross lipemia	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Whole blood	Ambient (preferred)	72 hours	
	Refrigerated	72 hours	

Clinical & Interpretive

Clinical Information

Sezary syndrome (SS) and mycosis fungoides (MF) are two distinct but intimately related T-cell lymphoproliferative disorders involving the skin and are commonly referred to as cutaneous T-cell lymphomas (CTCLs). SS is defined by the triad of erythroderma, generalized lymphadenopathy, and the presence of circulating cells with irregular nuclear features (Sezary cells). MF typically presents with slowly progressing patch and plaque lesions. Detection of neoplastic CD4-positive T cells in peripheral blood (>1000 cells/microliter) is essential to establish a diagnosis of SS. Disease staging and assessment of therapy response in CTCL require a quantitative assessment of peripheral blood involvement in absolute number of neoplastic cells (Sezary cells) per microliter. Flow cytometry is now considered the method of choice to estimate the number of Sezary cells in peripheral blood, largely replacing the less reproducible and time-consuming morphologic quantitation of atypical lymphocytes on a peripheral blood smear, proposed by the International Society for Cutaneous Lymphomas, and the cutaneous lymphoma task force of the European Organization of Research and Treatment of Cancer. Typically, Sezary cells are immunophenotypically distinct and they are clonal.

Reference Values

An interpretive report will be provided. This test will be processed as a laboratory consultation. An interpretation of the immunophenotypic findings and, if available, morphologic features will be provided by a board-certified hematopathologist for every case.

Interpretation

An immunophenotypically distinct T-cell population is suggestive of clonality when the subset exhibits a restricted T-cell receptor beta-chain constant (TRBC) staining pattern defined as either 1) >85% of TRBC1-positive events, 2) <15% TRBC1-positive events, or 3) homogenous TRBC1-dim expression. The immunophenotype of the distinct T-cell population, its percentage of total lymphocytes, and its percentage of total analyzed events will be reported. The test will be resulted as "No phenotypically aberrant T-cell population detected" if there is no specific immunophenotype that allows the detection of TRBC-restricted T cells.

Cautions

Correlation with clinical features is necessary for diagnosis of Sezary syndrome. This analysis can only describe a cell population with aberrant phenotype, but the significance of this finding in isolation is uncertain.

Clinical Reference

1. Horna P, Deaver DM, Qin D, et al. Quantitative flow cytometric identification of aberrant T cell clusters in erythrodermic cutaneous T cell lymphoma. Implications for staging and prognosis. J Clin Pathol. 2014;67(5):431-436
2. Berg H, Otteson GE, Corley H, et al. Flow cytometric evaluation of TRBC1 expression in tissue specimens and body fluids is a novel and specific method for assessment of T-cell clonality and diagnosis of T-cell neoplasms. Cytometry B Clin Cytom. 2021;100(3):361-369
3. Horna P, Shi M, Olteanu H, Johansson U. Emerging role of T-cell receptor constant beta chain-1 (TRBC1) expression in the flow cytometric diagnosis of T-cell malignancies. Int J Mol Sci. 2021;22(4):1817
4. Wilcox RA. Cutaneous T-cell lymphoma: 2016 update on diagnosis, risk-stratification, and management. Am J Hematol. 2016;91(1):152-165. doi:10.1002/ajh.24233
5. Horna P, Olteanu H, Jevremovic D, et al. Single-antibody evaluation of T-cell receptor beta constant chain monotypia by flow cytometry facilitates the diagnosis of T-cell large granular lymphocytic leukemia. Am J Clin Pathol. 2021;156(1):139-148
6. Horna P, Shi M, Jevremovic D, Craig FE, Comfere NI, Olteanu H. Utility of TRBC1 expression in the diagnosis of peripheral blood involvement by cutaneous T-cell lymphoma. J Invest Dermatol. 2021;141(4):821-829
7. Scarisbrick JJ, Hodak E, Bagot M, et al. Blood classification and blood response criteria in mycosis fungoides and Sezary

syndrome using flow cytometry: recommendations from the EORTC cutaneous lymphoma task force. Eur J Cancer. 2018;93:47-56

8. Illingworth A, Johansson U, Huang S, et al. International guidelines for the flow cytometric evaluation of peripheral blood for suspected Sézary syndrome or mycosis fungoides: Assay development/optimization, validation, and ongoing quality monitors. Cytometry B Clin Cytom. 2021;100(2):156-182

Performance

Method Description

Flow cytometry immunophenotyping of peripheral blood is performed using the following antibodies:
-Triage Panel: CD3, CD10, CD16, CD19, CD34, CD45, and kappa and lambda immunoglobulin light chains.
-Sezary Panel: CD2, CD3, CD4, CD5, CD7, CD8, CD26, CD45, and TRBC1. (Shi M, Jevremovic D, Otteson GE, Timm MM, Olteanu H, Horna P: Single antibody detection of T-cell receptor alpha-beta clonality by flow cytometry rapidly identifies mature T-cell neoplasms and monotypic small CD8-positive subsets of uncertain significance. Cytometry B Clin Cytom. 2020 Jan;98[1]:99-107)
-Possible additional panels performed per algorithmic approach.

PDF Report

No

Day(s) Performed

Monday through Saturday

Report Available

1 to 3 days

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

Fees & Codes

Fees

- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification

This test was developed using an analyte specific reagent. Its performance characteristics were determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and

Drug Administration.

CPT Code Information

88184-Flow cytometry; first cell surface, cytoplasmic or nuclear marker x 1
88185-Flow cytometry; additional cell surface, cytoplasmic or nuclear marker (each)
88188-Flow Cytometry Interpretation, 9 to15 markers (if appropriate)
88189-Flow Cytometry Interpretation, 16 or more markers (if appropriate)

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
SZDIA	Sezary Diagnostic Flow Cytometry, B	101118-8

Result ID	Test Result Name	Result LOINC® Value
CK126	Sezary Diagnostic	No LOINC Needed
CK127	Final Diagnosis	50398-7
CK128	Special Studies	30954-2
CK129	Microscopic Description	22635-7