Test Definition: SZMON

Sezary Monitoring Flow Cytometry, Blood

Overview

Useful For

Monitoring response to therapy in patients with previously diagnosed Sezary syndrome or mycosis fungoides

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 previously diagnosed Sezary syndrome or cutaneous T-cell lymphoma (CTCL) with peripheral blood involvement.

The panel is charged based on number of markers tested (FIRST for first marker, ADD1 for each additional marker).

Method Name

Immunophenotyping

NY State Available

Yes

Specimen

Specimen Type

Whole blood

Ordering Guidance

This test is for monitoring response to therapy in patients who have been diagnosed with Sezary syndrome or mycosis

fungoides. For patients with a clinical suspicion, but no diagnosis, of Sezary syndrome, order SZDIA / Sezary Diagnostic Flow Cytometry, Blood. A triage panel will also be performed to evaluate for and exclude monotypic B cells or increased blasts.

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)	4 days	
	Refrigerated	4 days	

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.

Interpretation

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.

An immunophenotypically distinct T-cell population is suggestive of clonality when the subset exhibits a restricted T-cell receptor beta-chain (TRBC) staining pattern defined as either greater than 85% of TRBC1-positive events, less than 15% TRBC1-positive events, or 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 and T-cell receptor beta-chain restriction, 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 Sezary 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:
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;98[1]:99-107)

PDF Report

No

Day(s) Performed

Monday through Saturday

Report Available

1 to 3 days

Specimen Retention Time

14 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Main Campus

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

SZMON	Sezary Monitoring Flow Cytometry, B	101117-0

Result ID	Test Result Name	Result LOINC® Value
CK130	Sezary Monitoring	No LOINC Needed
CK131	Final Diagnosis	50398-7
CK132	Special Studies	30954-2
CK133	Microscopic Description	22635-7