

**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**

Sezary panel is ordered in cases with previously diagnosed Sezary syndrome or cutaneous T-cell lymphoma (CTCL) with peripheral blood involvement. For cases without a previously confirmed diagnosis or previously performed immunophenotyping at our laboratory, the ordered test will be changed to SZDIA / Sezary Diagnostic Flow Cytometry, Blood, which includes a triage panel to exclude a B-cell lymphoproliferative disorder and a Sezary panel.

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).

**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 who do not have a diagnosis of Sezary syndrome, SZDIA / Sezary Diagnostic Flow Cytometry, Blood is the appropriate test to order.

### Specimen Required

#### Collection Container/Tube:

**Preferred:** Yellow top (ACD)

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

**Specimen Volume:** 6 mL

#### Collection Instructions:

1. Send in original tube. Do not transfer blood to other containers.
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 and Interpretive

### Clinical Information

Sezary syndrome is a leukemic form of cutaneous T-cell lymphoma (CTCL). By definition, it is associated with systemic skin involvement (erythroderma) and the presence of at least 1000/mcL of circulating cells with irregular nuclear features (Sezary cells). Morphologic assessment of the number of Sezary cells has been proven to have low reproducibility. Therefore, WHO/European Organization for Research and Treatment of Cancer (EORTC) classification of skin tumors adopted alternative methods to assess circulating T-cells in order to establish the diagnosis of Sezary syndrome. These include CD4:CD8 ratio of more than 10:1, and selective loss of CD7 and/or CD26 on 40% and 30% of the CD4-positive T-cell population, respectively. It is important to recognize that the later criteria (fulfilled by peripheral blood flow cytometry immunophenotyping) are relative, and not in direct correlation with

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absolute counts of Sezary cells defined by morphology.

### 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

Sezary cells typically show loss of CD7 and/or CD26. As loss of these markers is not completely sensitive or specific for Sezary cells, the WHO/European Organization for Research and Treatment of Cancer (EORTC) classification of skin tumors proposed cutoffs of 30% for CD26 loss and 40% for CD7 loss on CD4-positive T-cells, as diagnostic criteria for Sezary syndrome. In addition, CD4:CD8 ratio of greater than or equal to 10:1 in a gated T-cell population is also considered abnormal, and part of diagnostic algorithm for Sezary syndrome.

In mycosis fungoides staging studies the cutoffs are even less clearly defined. The clinical outcome was worse in patients with more than 5% of circulating lymphocytes showing Sezary-like morphology. However, flow cytometry immunophenotyping is deemed useful for relative quantification of these cells only if they can be separated by aberrant expression of other surface markers. In majority of cases, this cannot be accomplished to the proposed cutoff point (5% of circulating lymphocytes).

The test will be resulted as "No phenotypically aberrant T-cell population detected" if there is no specific phenotype that allows separation of potentially abnormal CD4-positive T-cells, loss of CD26 (and/or CD7) is present in less than 30% (40%), and CD4:CD8 ratio is less than 10:1. If any of the above aberrancies are present, the test will be resulted as "Phenotypically distinct T-cell population is detected" with a description of phenotype, percentage of total CD4-positive population and percentage of total analyzed events. In addition, the phenotype will be compared to that of any distinct T-cell population previously seen in the same patient by our laboratory.

### 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. Honra 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;67:431-436
2. Vaughan J, Harrington AM, Hari PN, et al: Immunophenotypic stability of Sezary cells by flow cytometry: usefulness of flow cytometry in assessing response to and guiding alemtuzumab therapy. *Am J Clin Pathol.* 2012 Mar;137(3):403-411
3. Kelemen K, Guitart J, Kuzel TM, et al: The usefulness of CD26 in flow cytometric analysis of peripheral blood in Sezary syndrome. *Am J Clin Pathol.* 2008 Jan;129(1):146-156
4. Wilcox RA. Cutaneous T-cell lymphoma: 2016 update on diagnosis, risk-stratification, and management. *Am J Hematol.* 2016;91:152-165. doi: 10.1002/ajh.24233
5. Olsen E, Vonderheid E, Pimpinelli N, et al: Revisions to the staging and classification of mycosis fungoides and Sezary syndrome: a proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous lymphoma task force of the European Organization of Research and Treatment of Cancer (EORTC). *Blood.* 2007 Sep 15;110(6):1713-1722
6. Willemze R, Jaffe ES, Burg G, et al: WHO-EORTC classification for cutaneous lymphomas. *Blood.* 2005;105:3768-3785

## 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 Jan;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

Rochester

## Fees and 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 Regional Manager. 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

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Test ID	Test Order Name	Order LOINC Value
SZMON	Sezary Monitoring Flow Cytometry, B	In Process

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