

## 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.

[For patients with a clinical suspicion of Sezary syndrome, but no previously confirmed diagnosis or immunophenotyping performed in our laboratory, 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.](#)

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.

### 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. 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 (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 resultated 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

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 |
|---------|-------------------------------------|--------------------|
| 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             |