MYCOSIS FUNGOIDES OR SÉZARY SYNDROME DATA SHEET (ICD10 C84.0 and C84.1)

UICC TNM 9th EDITION STAGING SUMMARY

MYCOSIS FUNGOIDES OR SÉZARY SYNDROME

Prognostic Group	**T stage**	**N stage**	**M stage**	**Peripheral Blood Involvement**
IA	T1	N0	M0	B0, B1
IB	T2	N0	M0	B0, B1
IIA	T1, T2	N1, N2	M0	B0, B1
IIB	T3	N0, N1, N2	M0	B0, B1
III	T4	N0, N1, N2	M0	B0, B1
IIIA	T4	N0, N1, N2	M0	B0
IIIB	T4	N0, N1, N2	M0	B1
IVA1	Any T	N0, N1, N2	M0	B2
IVA2	Any T	N3	M0	Any B
IVB	Any T	Any N	M1	Any B

Notes:
This is a prognostic group as the prognostic group is determined by the absence or presence of peripheral blood involvement.
Clone status is not required to assign a prognostic group.

Mycosis Fungoides or Sézary syndrome

T – Primary Tumour	
T1	Limited patches. papules, and/or plaques covering < 10% of the skin surface **T1a** – Patch only **T1b** – Plaque/papule ± patch
T2	Patches, papules, or plaques covering ≥ 10% of the skin surface **T2a** – Patch only **T2b** – Plaque/papule ± patch
T3	One or more tumours (≥1 cm in diameter)
T4	Confluence of erythema covering ≥80% of the body surface

N – Regional Lymph Nodes	
NX	Clinically abnormal peripheral lymph nodes, no histologic confirmation
N0	No clinically abnormal peripheral lymph nodes*, biopsy not required
N1	Clinically abnormal peripheral lymph nodes; histopathology Dutch grade 1 or National Cancer Institute (NCI) LN0-2 **N1a** – Clone negative** or equivocal **N1b** – Clone positive** and identical to skin
N2	Clinically abnormal peripheral lymph nodes; histopathology Dutch grade 2 or NCI LN3 **N2a** - Clone negative** or equivocal **N2b** - Clone positive** and identical to skin
N3	Clinically abnormal peripheral lymph nodes; histopathology Dutch grades 3-4 or NCI LN4 **N3a** - Clone negative** or equivocal **N3b** - Clone positive** and identical to skin

Note

* A lymph node (LN) of >1.5 cm in the longest diameter (LDi) is considered abnormal, especially if accompanied by other features of concern (firm, rubbery and/or fixed) but should be confirmed by imaging. Central nodes, which generally are not amenable to pathological assessment, currently are not considered in the nodal classification unless used to establish N3 histopathologically.

** A T cell clone is defined by polymerase chain reaction (PCR) or Southern blot analysis of the TCR genes.

For more detailed information regarding TNM staging, please see pages 234-236 of the UICC TNM 9 book

M – Distant Metastasis	
M0	No visceral organ involvement*
M1a	Bone marrow only **M1a1** - Clone negative** or equivocal **M1a2** – Clone positive** and identical to skin
M1b	Non-bone marrow visceral involvement* with or without bone marrow involvement **M1a1** - Clone negative** or equivocal **M1a2** – Clone positive** and identical to skin
* For viscera, spleen and liver may be diagnosed by imaging criteria ** A T cell clone is defined by polymerase chain reaction (PCR) or Southern blot analysis of the TCR genes.	

Histopathologic staging of lymph nodes in Mycosis Fungoides and Sézary syndrome

EORTC classification	**Dutch system**	**NCI-VA classification**
N1	Grade 1: Dermatopathic lymphadenopathy (DL)	LN0: No atypical lymphocytes
		LN1: Occasional and isolated Atypical Lymphocytes (not arranged in clusters)
		LN2: Many atypical lymphocytes or lymphocytes in 3-6 cell clusters
N2	Grade 2: DL; early involvement b MF (presence of cerebriform nuclei <7.5 μm)	LN3: Aggregates of atypical lymphocytes; nodal architecture preserved.
N3	Grade 3: partial effacement of lymph node architecture; many atypical cerebriform architecture by atypical lymphocytes mononuclear cells	LN4: Partial/complete effacement of nodal or frankly neoplastic cells.
	Grade 4: complete effacement	

Peripheral Blood Involvement (B)

B Category	**B Criteria**
B0	Absence of significant blood involvement: ≤250/μL of CD4+/CD26- or CD4+/CD7- cells **B0a** – Clone negative** **B0b** – Clone positive**
B1	Low blood tumour burden: >250/μL but ≤1000/μL of CD4+/CD26- or CD4+/CD7- cells **B1a** – Clone negative** **B1b** – Clone positive**
B2	High blood tumour burden: >1000/μL of CD4+/CD26- or CD4+/CD7- cells or other aberrant population of lymphocytes identified by flow cytometry.
** A T cell clone is defined by polymerase chain reaction (PCR) or Southern blot analysis of the TCR genes.	