

QoL-P-03

Quality of life in patients with Mycosis fungoides and Sezary Syndrome:
a systematic review of the literature

Introduction

Cutaneous T-cell Lymphoma’s (CTCL) are a rare, heterogeneous group of T-cell lymphomas that primarily manifest in the skin. Mycosis fungoides (MF) and Sezary syndrome (SS) are considered the classic types of CTCL. The diverse manifestation of CTCL results in a wide range of symptoms with a possible mild to severe impact on Quality of Life (QoL) depending on the disease stage. Previous studies on QoL in CTCL patients report diverse patient populations and use many different QoL instruments. In the current literature, a clear overview on the influence of the different stages of disease (early MF, late-stage MF/SS or total group) on the QoL is lacking.

Method

A systematic search of the literature was conducted using the Pubmed, Embase, PsycINFO and Web of Science databases. Studies were included if they described QoL in patients with MF and SS retrieved by standardized instruments or qualitative interviews.

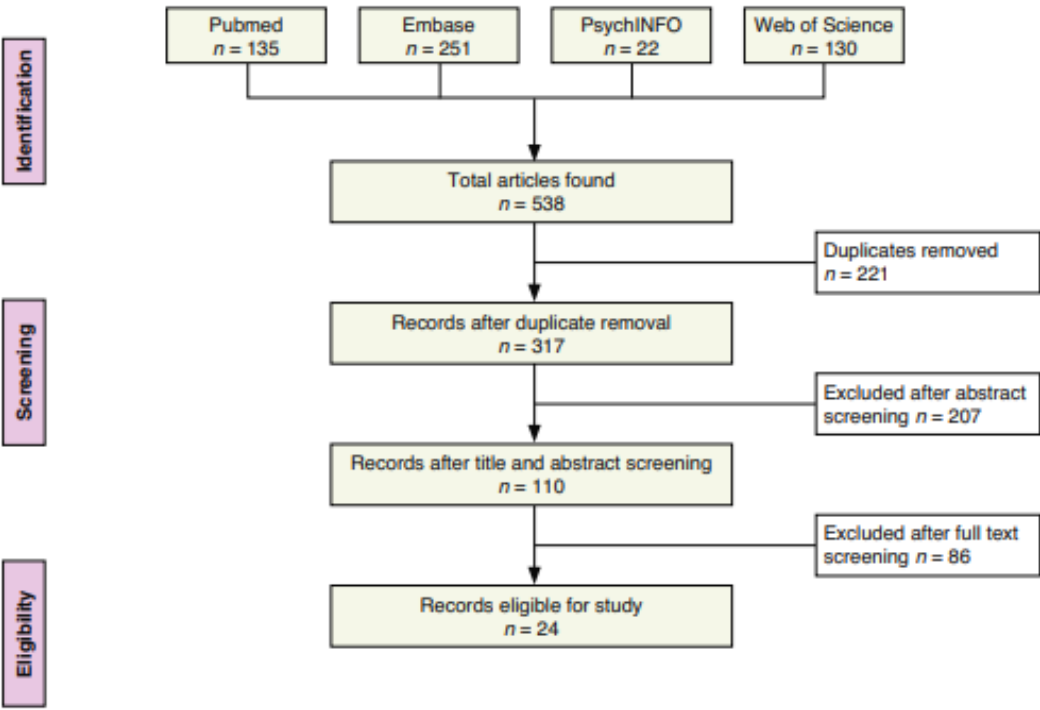


Table 1 Study characteristics of included studies

Author	Year	Country	n	Age (mean or median)	Diagnosis	Stage	Reported stage	Skin Specific QoL	Generic QoL	Cancer-specific QoL	Pruritus	Other
Quantitative												
Ayyalaraju et al ¹⁰	2003	UK, USA	51	*	MF	Hospitalized	Mixed	DLQI				
Booken et al ¹¹	2011	Germany	5	µ 61 (51–68)	SS/erythrodermic MF	MF IIIB, SS IVA	Late	DLQI			VAS	
Bouwhuis et al ¹²	2003	USA	20	*	SS		Late		SF-36			
Demierre et al ¹³	2007	USA	11	M 62 (39–80)	MF/SS	IB-IV	Mixed	SD-29		FG-V4		
Demierre et al ⁵	2006	USA	630	M 57	MF/SS	IA-IV	Mixed					Self-made
Demierre et al ¹⁴	2005	USA	22	µ 63.1 (32–81)	MF	I-IIA, II-IVB	Early and late	SD-29		FG-V4		
Duvic et al ¹⁵	2001	USA	94	M 64 (27–89)	CTCL	IIIB-IVB	Late			Spitzer		
Duvic et al ¹⁶	2002	USA	71	µ 61	MF/SS	IB-III	Mixed			FG-V3	VAS	
Engin et al ⁶	2020	Turkey	52	µ 46	MF	IA-IVB	Mixed		SF-36			BDI, BAI
Herbosa et al ¹⁷	2020	USA	115	M Early stage 64.2 (59.2–72.2) M Late stage 67.1 (47.0–71.9)	MF/SS	IIIB-IVB	Early and late	SD-29	SF-36, HUI-3		VAS	
Holahan et al ¹⁸	2018	USA	105	µ 61.07	MF/SS	I-IV	Mixed	SD-16	EQ5D			
Illidge et al ¹⁹	2013	UK	36	M 65 (38–83)	MF	IB-IVA	Mixed	SD-29		qlq-c30	VAS	
Jennings et al ²⁰	2019	USA	8	(37–81)	MF	*	Mixed	DLQI			VAS	
Molloy et al ²¹	2020	Multinational	238	M 60 (49–70)	MF/SS	IA-IVB	Mixed	SD-29				
Porkert et al ²²	2018	Austria	55	µ 63.8(±13.27)	CTCL	IA-IIA, IIB-IVB	Early and late	SD-29				IPO-R
Quaglino et al ²³	2020	Multinational	56	M 57 (5–97)	MF	IA-IIA	Early	SD-29				
Sampogna et al ²⁴	2009	Italy	95	µ 56 (±18)	MF/SS	IA-III	Early, late, and mixed	SD-29		qlq-c30		
Semenov et al ²⁵	2020	USA	67	µ 65 (±12.8)	MF/SS	IA-IVB	Early and late		HUI-3			
Steinke et al ²⁶	2018	Multinational	15	µ 51.1 (± 18.2)	MF/SS	*	Mixed	DLQI			NRS Itchy-QoL	
Wright et al ²⁷	2013	UK	100	µ 57.9 (± 12.9)	MF/SS	I-IV	Early, late, and mixed	SD-29			VAS	
Qualitative												
Beynon et al ²⁸	2015	UK	19	41-83	CTCL	IAB, IIB, III, IVA	Mixed					Semi-structured
Bhat et al ²⁹	2020	USA	18	M 62 (IQR18)	MF/SS	IIB-IV	Early					Interview
Demierre et al ³⁰	2011	UK, USA	19	M 64 (25–80)	MF/SS	IA-IVA	Mixed	SD-16				Interview
Selman et al ³¹	2015	UK	14	36–85	MF/SS	IB-IVB	Mixed					Semi-structured

n, number of patients; *, missing; ±, SD; (), range; µ, mean; M, median; MF, Mycosis fungoides; SS, Sézary syndrome; DLQI, Dermatology Life Quality Index; SD-29/16, Skindex-29/16; SF-36, Shortt Form Health Survey, HUI-3, Health Utility Index; EQ5D, EuroQoL-5D; FG-V4/3, Functional Assessment of Cancer Therapy – General Version 4/3; QLQ-C30, Core Quality of Life questionnaire; VAS, Visual Analogue Score; SD, Pruritus-SD Itch scale; Itchy-QoL, Itchy Quality of Life; BDI, Back Depression Inventory; BAI, Beck Anxiety Inventory; IPO-R, Revised Illness Perception Questionnaire.

Results

In total, 24 studies were included using 18 different questionnaires to report on dermatology-specific, cancer-specific and generic QoL. The effect on QoL was found to be greater in patients with late-stage disease as compared to early stage disease, with significant impairments on functional, emotional and physical domains. Nonetheless, even in patients with limited disease, QoL was mildly to moderately affected. Overall, pruritus was the most frequent reported and most bothersome symptom. Significant influence of the disease on daily life activities were found, not only in patients but also on caregivers and family.

Conclusion

This broad, structured overview on QoL in MF and SS patients underlines the influencee of disease stage on QoL, and therefore, recommends future studies to distinguish between disease stages when reporting results.

Furthermore, this overview can inform clinicians in clinical practice by creating awareness of QoL deficits according to disease stage.

References

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Conflicts of interest

R. Willemze: Participation on a Data Safety Monitoring Board or Advisory Board; Helsinn Healthcare S.A
M.H. Vermeer: Payment or honoraria for lectures, presentations, speakers bureaus, manuscript writing or educational events and Participation on a Data Safety Monitoring Board or Advisory Board; Kyowa