The PROCLIPI international registry of early-stage mycosis fungoides identifies substantial diagnostic delay in most patients


BACKGROUND
Survival in mycosis fungoides (MF) is varied and may be poor. The PROCLIPI (PROspective Cutaneous Lymphoma International Prognostic Index) study is a web-based data collection system for early-stage MF with legal data-sharing agreements permitting international collaboration in a rare cancer with complex pathology. Clinicopathological data must be 100% complete and in-built intelligence in the database system ensures accurate staging.

OBJECTIVES
To develop a prognostic index for MF.

METHODS
Predefined datasets for clinical, haematological, radiological, immunohistochemical, genotypic, treatment and quality of life are collected at first diagnosis of MF and annually to test against survival. Biobanked tissue samples are recorded within a Federated Biobank for translational studies.

RESULTS
In total, 430 patients were enrolled from 29 centres in 15 countries spanning five continents. Altogether, 348 were confirmed as having early-stage MF at
central review. The majority had classical MF (81.6%) with a CD4 phenotype (88.2%). Folliculotropic MF was diagnosed in 17.8%. Most presented with stage I (IA: 49.4%; IB: 42.8%), but 7.8% presented with enlarged lymph nodes (stage IIA). A diagnostic delay between first symptom development and initial diagnosis was frequent [85.6%; median delay 36 months (interquartile range 12-90)]. This highlights the difficulties in accurate diagnosis, which includes lack of a singular diagnostic test for MF.

CONCLUSIONS
This confirmed early-stage MF cohort is being followed-up to identify prognostic factors, which may allow better management and improve survival by identifying patients at risk of disease progression. This study design is a useful model for collaboration in other rare diseases, especially where pathological diagnosis can be complex.