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8.3 Disease patterns in Danish Juvenile Dermatomyositis patients

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Purpose

- Retrospective assessment of disease characteristics and outcome for the Danish cohort of Juvenile Dermatomyositis (JDM) patients, 1977 – 2005.

- Evaluation of the Myositis disease activity assessment tool (MYOACT) and Myositis intention to treat activity index (MITAX) as prognostic tools.

Methods

Hospital records from Danish JDM patients (1977 – 2005) were reviewed. The parameters of the MYOACT and MITAX were used for the database.

Results

53 patients were classified as JDM. The female:male ratio was 2:1, the mean age at disease onset was 7.1 years and the mean disease duration was 3.6 years. Most frequent symptoms at disease onset are displayed in Table 1.

At the 5-years follow-up 34% were in remission, 30% had ongoing disease and disease- or treatment-induced damage was present in 36%. In the total follow-up period (2–30 years) 3 patients (6%) had died, 68% were in full remission, 13% had ongoing disease and 13% had unknown status.

Conclusion

- Most patients had a favourable outcome; however irreversible damage was found in 36% at 5-years follow-up

- Baseline predictors of unfavourable disease outcome could not be identified

- Due to frequently missing chart data MYOACT and MITAX could not be used as a scoring tool in this retrospective set-up

- A clinical long-term follow-up study is warranted and now carried out by the authors.

Table 1: Most frequent symptoms at disease onset

Symptom	% of patients
Proximal muscle weakness	81
Fatigue	74
Erythema	74
Gottrons papules	70
Heliotrope	57
Arthralgia	41
Fever	32
Arthritis	30
Weight loss	30