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Trends in Medication Usage in Patients with Juvenile Dermatomyositis

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SESSION INFORMATION

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Session Type: ACR Poster Session B

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Background/Purpose: Juvenile dermatomyositis (JDM) is a systemic autoimmune disease with characteristic rashes and chronic muscle inflammation. Because of its rarity, most therapeutic choices are based on small trials or retrospective series. The purpose of this study was to evaluate changes in treatment over time, and factors associated with medication choices in JDM.

Methods: We performed a retrospective review of therapies received by 334 patients with probable or definite JDM enrolled in the Childhood Myositis Heterogeneity Study. Patients were diagnosed from November 1965 to September 2014 (median August 1994). We evaluated the number and type of medications received and their durations, including differences by year of diagnosis, onset severity, and myositis autoantibodies (MSAs). Logistic regression and ROC analysis determined 1997 as a cut point for increasing usage of drugs other than prednisone.

Results: The median follow-up duration was 46 months and treatment duration was 33 months. Oral prednisone (PRED) was the primary therapy used by 99.4% of JDM patients, but was used as monotherapy in only 49% of patients diagnosed after 1997 vs. 85% before 1997 (P<0.001). Other medications were used more frequently in patients diagnosed after 1997 vs. before, including methotrexate (MTX) (95% vs. 60%), Intravenous Immunoglobulin (IVIG) (64% vs. 25%), hydroxychloroquine (HCQ) (70% vs. 37%), other DMARDs (41% vs. 14%) and cytotoxics/biologics

(28% vs. 6%, P<0.001 for all). Patients diagnosed after 1997 had a greater number of drug trials per year (median 2.3 vs. 1.0) and a longer percentage of follow-up time on treatment compared to patients diagnosed before 1997. The median daily maximum PRED dose was 2.0 mg/kg/d [IQR 1.2-2.0], and did not differ by onset severity or MSAs. However, the median time to half the initial PRED dose was shorter in patients diagnosed after vs. before 1997 (11 vs. 22 months, P<0.01). The median time to discontinuation of PRED was 47 months [IQ 24-104] and for MTX was 50 months [IQ 24-86] by Kaplan-Meier analysis. Thirty-seven percent of patients discontinued all medications at last follow-up, with a median time to discontinuation of 85 months. There were no significant differences in time to discontinuation of PRED, MTX, or other medications by onset severity or MSAs, but there was a longer time to discontinuation for IVMP, MTX and IVIG in patients diagnosed after 1997 (P<0.01). We examined factors for use of IVMP, MTX, IVIG, and HCQ by multiple logistic regression. All drugs had greater usage in patients diagnosed after 1997 (OR 2.2-13.4). IVMP was more frequently used in patients with severe onset (OR 3.0), with anti-p155/140 and anti-MJ Abs (OR 2.7 and 2.5), and with a higher total initial symptom score (OR 21.9). HCQ was used more commonly in patients with anti-p155/140 Abs (OR 2.2). MTX was more frequently used in older patients at diagnosis (OR 1.1).

Conclusion: PRED is the mainstay of therapy in JDM. There is increasing use of MTX, IVIG and other drugs/biologics in combination after 1997, and PRED was reduced faster for this group compared with patients diagnosed before 1997. Diagnosis year, onset severity, age at diagnosis, and MSAs were associated with specific medication usage in JDM.

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