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MEDIUM AND LONG TERM FOLLOW UP IN A MULTICENTER COHORT OF PATIENTS WITH JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHIES IN HUNGARY

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Introduction: Dermatomyositis(DM)/polymyositis(PM), belongs to the group of the idiopathic inflammatory myopathies. It is characterized by bimodal pattern of age-specific incidence of rates, with peaks in age group from 5 to 14 years (juvenile DM/PM) and from 45 to 64 years (adult DM/PM). The aim of this study is to evaluate the clinical characteristics of patients with juvenile dermatomyositis and –polymyositis in Hungary. We have recently compiled a questionnaire to summarize clinical experiences of diagnosing and managing juvenile DM patients. We searched relation between the initial treatment and the following clinical course. Methods: We analyzed the medical records of 41 patients with juvenile dermatomyositis/polymyositis. The patients were divided in 3 groups based on their clinical course: monophasic, polycyclic and chronic. Results: The prevalence of polycyclic and monophasic subtypes of the disease were found similar (15/41 and 22/41). All children were given glucocorticoids. 11/41 (27%) patients were resistant to glucocorticoids as a first-line agent, and they were treated with cyclosporin A. In this group less relapses of the disease were observed. During the clinical course 21/41 (51%) children were given second-line immunosuppressive agents. Finally, 35/41 patients achieved remission, however, 4/35 patients still have to take low-dose glucocorticoid therapy to maintain remission. 6 patients' cutaneous symptoms are persistent. Discussion: the authors compare their data of juvenile patients with the data of the relevant literature and to their experience with the management of adult DM patients.