# Liposteroid therapy for juvenile and adult dermatomyositis: efficacy and side effects

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Juvenile dermatomyositis (JDM) is a multisystem disease of uncertain origin that is defined by chronic inflammation of striated muscle and the skin (1). Similarities and differences between JDM and adult dermatomyositis (ADM) have been reported (1). We know that persistent muscle impairment, despite immunosuppressive therapy, is a common symptom for both patients with JDM and ADM. However, there is less information regarding the pathophysiology of JDM compared with ADM.

In patients with ADM, immunosuppressive therapy, including glucocorticoids, has suppressive effects on the gene expression of the inflammatory pathways in skeletal muscle, but it also changes the expression of genes involved in skeletal muscle tissue remodeling, which may negatively affect muscle regeneration and growth (2). The altered gene expression has been associated with lipid uptake and lipolysis in response to immunosuppressive therapy, which may also have a negative effect on muscle performance (2). It has been suggested that for patients with ADM, muscle impairment may persist, while the inflammation associated with the myositis improves after glucocorticoid therapy. The same pathophysiology may apply for JDM. Furthermore, in JDM, this mechanism may present not only as persistent muscle impairment, but also as persistent skin ulcers, despite immunosuppressive therapy (3,4). This is because a relationship between dermatitis and lipid metabolism has been reported (5). For both ADM and JDM, muscle biopsies reveal signs of inflammation, including infiltrating macrophages in the muscle fibers (6,7), whereas activated macrophages are present in skin ulcers with calcinosis in JDM (8).

Liposteroid [dexamethasone palmitate (DP)] is a lipid emulsion containing dexamethasone, which was developed in Japan (9). This agent provides greater efficacy, and much less risk for systemic adverse effects, than dexamethasone, because the lipid emulsion is easily taken up by phagocytes (10). When the same dose of DP or dexamethasone sodium phosphate (DSP) is administrated, the uptake of DP is 8-time higher, it is hydrolyzed slowly, and the anti-inflammatory effect on macrophages is 5- to 6-time higher than DSP (9,10). In addition, DP has fewer side effects, for example, it has little inhibitory effect on the hypothalamus-pituitary-adrenal axis compared with DSP (9).

Japanese physicians have used liposteroid therapy to treat rheumatoid arthritis (11). Recently, Japanese researchers reported the clinical efficacy and utility of liposteroid in the treatment of diseases associated with macrophage activation, including hemophagocytic lymphohistiocytosis (12), pulmonary hemosiderosis (13), and graft-versus-host disease (14). There are no reports regarding the clinical utility of liposteroid for the treatment of JDM and ADM. However, we reported successful liposteroid therapy of JDM-associated macrophage activation syndrome (15). This was the first case to show the efficacy, with no adverse events, in high-dose liposteroid therapy for JDM-associated

macrophage activation syndrome. Although investigation of the effects of the lipid emulsion in liposteroid on lipid metabolism needs to be performed, and case reports regarding the use of liposteroid therapy for treatment of JDM and ADM need to be accumulated in order to confirm the clinical utility of liposteroid therapy for these conditions, there is some evidence that this agent may be effective for reducing side effects in patients with JDM and ADM, especially the muscle impairment and skin ulcers that persist despite immunosuppressive therapy.

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## Footnote

*Conflicts of Interest:* The author has no conflicts of interest to declare.

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