

# Juvenile Dermatomyositis and Extensive Calcinosis. Treatment With Methylprednisolone and Methotrexate

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Juvenile dermatomyositis (JDM) is considered a multisystemic disease of uncertain etiology. The clinical manifestation is a non-suppurative inflammation of the striated muscle, gastrointestinal tract, and skin. Dystrophic calcifications are present in 30%-70% of children with JDM.

The clinical case we are presenting is a 4 years old female with diagnosis of JDM in accordance to the Bohan and Peters criteria (very early presentation age) with extensive calcinosis, classified as functional class III, without being able to sit down or flex her knees. She was treated with IV methylprednisolone (MPS) bolus every 14 days and oral methotrexate, with improvement of her clinical condition. Even though calcinosis is a frequent finding in connective tissue disease and can cause severe disability, there are no treatment protocols at this time. The simultaneous use of IV MPS and oral methotrexate allows for a faster control of the disease, improvement in muscular force, reduction of erythema and regression of the calcinosis without important collateral effects.

**Key words:** Dermatomyositis. Calcinosis. Methylprednisolone.

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### Dermatomiositis juvenil y calcinosis extensa. Tratamiento con metilprednisolona y metotrexato

La dermatomiositis juvenil (DMJ) es una enfermedad multisistémica de etiología incierta, que resulta en una inflamación crónica no supurativa del músculo estriado,

la piel y el tracto gastrointestinal. Las calcificaciones distróficas ocurren en un 30-70% de los niños con DMJ. Presentamos el caso de una paciente de 4 años de edad, con diagnóstico de DMJ según criterios de Bohan y Peter, en una edad muy temprana de presentación, con calcinosis extensas que le impedían sentarse, sin flexión de articulación de rodillas, con clase funcional 3. Recibió tratamiento con pulsos intravenosos de metilprednisolona cada 14 días, además de metotrexato vía oral, con mejoría clínica.

A pesar de que la calcinosis es frecuente en enfermedades del tejido conectivo y puede llevar a discapacidad severa, no se han desarrollado protocolos terapéuticos para su manejo. El uso simultáneo de metilprednisolona y metotrexato permite un control más rápido de la enfermedad, con mejoría en la fuerza muscular y el eritema y regresión de las calcinosis, sin efectos colaterales importantes.

**Palabras clave:** Dermatomiositis. Calcinosis. Metilprednisolona.

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### Introduction

Juvenile dermatomyositis (JDM) is a multisystemic disease of unknown etiology that leads to a chronic, non supurative inflammation of the striated muscle, skin and gastrointestinal tract. It is characterized in its early stages by a small vessel vasculitis, and in its latter stages by the development of calcinosis.<sup>1</sup>

The diagnosis of JDM is established based on the criteria proposed by Bohan et al,<sup>2</sup> in the presence of a typical skin eruption (Gottron's papules, heliotrope erythema), as well as 3 of the following criteria: symmetric proximal muscle weakness, electromyographic of an inflammatory myopathy, an increase in muscle enzymes, and inflammatory myositis in the muscle biopsy.

Pathologic calcification of soft tissue, similar to those that occur on calcified heart valves, are a complication of some diseases of connective tissue which increase both morbidity and mortality.<sup>3</sup> In contrast with dermatomyositis in adults,

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Manuscript received September 6, 2007; accepted for publication November 28, 2007.

in which calcifications are uncommon, an estimated 20%-40% of children with JDM have calcified deposits.<sup>4-6</sup> In JDM, calcifications occur more frequently in the anatomical areas which are normally exposed to micro trauma but not mineralized, such as elbows and knees.<sup>4</sup>

We present the case of a patient with JDM who presented calcinosis and muscle contraction as part of her progression.

### Clinical Case

A 4 year, 5 month old girl was diagnosed with systemic lupus erythematosus at age 2 due to facia erythema, fotosensitivity, weight loss, anorexia, fever, and a reduction in muscle strength, and was treated with prednisone at different doses in another hospital. After 2 years of progression, she was seen at our hospital with generalized facial edema, heliotrope erythema, Gottron's papules, skin rigidity of the thorax, arms, abdomen, thighs, and legs. Calcinosis of different sizes was seen on the arms, abdomen, buttocks, and thighs. Her muscle mass was painful upon

palpation and she had a muscle strength of 2/5. She had an important limitation of flexion and extension of the shoulders in 5°, flexion of the elbows in 90°, and extension of 80°, the hips and knees were fixed at 0° flexion, and was incapable of sitting. Anti-DNA antibodies, antinuclear antibodies, anti-Ro, anti-La, anti-Sm, and anti-RNP antibodies were negative. The blood analysis showed: LDH, 1383 U; CK, 56 U; ESR, 55 mm/first hour. The electromyography showed myopathic changes. The x-rays showed generalized calcinosis in the form of a thoracic shell, affecting the abdomen and extremities (Figures, A and B). Treatment with pulse methylprednisolone (3 doses of 30 mg/kg/day), methotrexate (28.8 mg/m<sup>2</sup>/week), folic acid, naproxen (20 mg/kg/day) and prednisone at a dose 60 mg/day for 6 weeks, with a descending dosage. She received methylprednisolone pulse therapy every 2 weeks from November 2006 up to the time of this writing, completing a total of 20 doses. Currently, the patient has presented notable improvement, with a significant regression of calcinosis (Figures, C and D), with discreet facial erythema and a muscle strength of 4/5. She persists

**Figure.** A and B: generalized calcinosis observable in simple x-rays of the abdomen, pelvis, and lower extremities upon diagnosis. C and D: the same patient, with generalized calcinosis in remission after treatment with intravenous methylprednisolone.

with joint contracture due to a lack of movement, with thorax flexion range of motion arches of 60°; hips, 30°; knees, 40°; elbows, flexion of 150° and extension of 50°, being able to sit at a 90° angle with help. She is currently in class II functional stage.

## Discussion

The prognosis of children with JDM has improved since the introduction of steroid therapy; however, there is still considerable morbidity. In particular, the deposit of calcium in the skin (calcinosis cutis), surrounding the joints (circumspect calcinosis), and the intermuscular fascia (universal calcinosis) can lead to more incapacity in the long term than the myopathic inflammation itself.<sup>6</sup> In the present case, JDM was diagnosed according to the criteria proposed by Bohan and Peter, with a very early age of presentation.

Mild calcinosis cutis frequently disappears with treatment, but more severe cases can cause severe chronic pain, persistent ulcers and ulcer infection, abscess formation, joint contraction, fever and systemic complications, even death.<sup>7</sup> Different therapeutic strategies have been published for calcinosis, such as the use of biphosphonates (alendronate), aluminum hydroxide, probenecid, warfarin, colchicine, minocycline, salicylate, diltiazem, surgery, laser therapy with carbon dioxide, with different results.<sup>8-10</sup>

There is evidence that early and aggressive treatment of JDM with intravenous methylprednisolone pulse therapy can reduce the incidence or the severity of calcinosis as well as improve the functional outcome.<sup>7,11</sup> The simultaneous use of methylprednisolone and methotrexate allows for a faster control of the disease and an early suspension of oral steroids, with a reduced mortality,

something that was also seen in our patient, with a notable improvement in muscle strength and erythema and regression of calcinosis as evidenced by the improvement in the range of movement of the different affected joints, without collateral effects.<sup>7,11-13</sup> Long term studies with a larger number of patients will provide more information on the clinical progression and the possible therapeutic effects as applied to our patient.

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