

## Pulse Therapy in Scleroderma

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Scleroderma is a generalized, multisystem disease in which characteristic vascular alterations are seen together with severe fibrosis of skin, synovium and certain internal organs, chiefly the intestinal tract, lung, heart and kidneys. An autoimmune etiology has been suggested by the clinical association with other diseases of presumed autoimmune etiology. The presence of Raynaud's phenomenon in 95% of scleroderma patients called attention to a possible vascular pathogenesis of the disease(1).

The disease is exceptionally rare in childhood (only 57 cases have been reported in the world literature) and only five cases could be culled from the records of some 15 years in the Royal Hospital for sick children, Glasgow(2). It affects girls more frequently than boys (4:1) and may begin at any time during childhood. There is no familial predisposition(3,4).

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Therapy in scleroderma has been generally discouraging. Pasricha has reported success with pulse therapy in certain skin disorders like pemphigus. Reiter's disease pyoderma gangrenosa and Darriers disease. One case of systemic sclerosis in an adult has also been successfully treated by the same author with dexamethasone pulses(5). This prompted us to give a trial with intermittent high doses of dexamethasone and cyclophosphamide to our patient who showed significant reversal of all the clinical manifestations without any side effects.

### Case Report

Mamta, 7<sup>1/2</sup> years daughter of a serving soldier, weighing 15 kg was admitted to our hospital on 28 June, 1992 with the history of inability to sit and walk since last 6 months. She had been ill for the last 1<sup>1/2</sup> yrs prior to hospitalization. The disease had been gradually progressive. On examination she had glistening hide bound skin all over the body including the trunk and face. The lower eye lids could not be everted. Mouth opening was restricted to 1.2 cm only. There were irregular multiple pigmented and depigmented patches distributed all over the body. There were contractures of both knees, ankles, elbows, wrists and small joints of the hands with marked restriction of movements. She had multiple ulcers on both legs and feet. There was no history of dysphagia. Raynaud's phenomenon was positive. Her spleen was palpable 2 cm below the left costal margin. Heart, lungs and neurological examination were essentially normal.

Her blood counts, liver function tests, renal functions, serum calcium, phosphorus, alkaline phosphatase, X-ray chest, barium swallow and ECG were within normal limits. Rheumatoid factor, antinuclear antibody and LE cell phenomenon were negative.

X-ray elbow and wrist did not reveal any calcinosis.

Skin biopsy showed atrophic dermal appendages. The collagen bundles in reticular dermis were thickened and appeared homogenous deep eosinophilic typical of progressive systemic sclerosis.

The child was treated with dexamethasone 2 mg/kg/day given in 100 ml of 5% of dextrose intravenously over a period of 30-60 minutes, on three consecutive days in a month. Injection cyclophosphamide was added to dexamethasone in dose of 2.5 mg/kg on the first day of the pulse. Such eight pulses were given. She was also given physiotherapy simultaneously.

The child showed remarkable improvement with this pulse therapy. Her general outlook improved tremendously. She gained weight by 2 Kg. The Raynauds phenomenon disappeared after two pulses. Mouth opening increased to 2.5 cm. There was palpable loosening of the skin and significant reduction in contractures alongwith improvement in pigmentary changes of skin. All the ulcers healed after 3 pulses. Spleen also regressed and was no longer palpable at the time of discharge. She was able to sit and walk independently. Repeat skin biopsy showed epidermis to be unremarkable. Reticular dermis showed less pronounced vascular changes and nonhomogenization of papillary dermis suggesting remarkable improvement as compared to previous biopsy report. The patient was discharged in February 93 as the father had to proceed on pension, with the advice to continue pulse therapy at the nearest hospital.

### Discussion

The high dose steroid pulse therapy using 1000 mg of methyl prednisolone was used with success in many dermatological

conditions. To reduce the dose of steroids without affecting the efficacy of the regime, cyclophosphamide was added and proved quite effective(5). The pulse therapy has distinct advantage over conventional daily dose therapy. Firstly, pulse therapy used often produces therapeutic response in cases where conventional daily doses have minimal effect. Secondly, this pulse therapy does not produce any of the well known side effects of prolonged use of corticosteroid-cyclophosphamide.

To the best of our knowledge this is the first case of progressive systemic sclerosis in a child, who has been treated successfully with the pulse therapy and the results obtained have been certainly more than what we anticipated. Not only was the progress of the disease arrested but the histopathological changes also showed significant reversal. It will however, be necessary to follow up this patient and also to try this method for other cases as well to see if all of them will have similar improvement.

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