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Treatment of dermatomyositis in childhood

VICTOR DUBOWITZ

From the Department of Paediatrics and Neonatal Medicine, Hammersmith Hospital, London

Dubowitz, V. (1976). *Archives of Disease in Childhood*, **51**, 494. **Treatment of dermatomyositis in childhood.** Analysis of the response to corticosteroid therapy in a personal series of 8 consecutive cases of dermatomyositis in childhood shows that there are advantages in a moderate dosage, short-term treatment schedule, with gradual tapering of the dosage as soon as there is clinical improvement without waiting for full remission, and in trying to stop steroid therapy within six months rather than following the more prolonged regimen currently still in vogue. Clinical response is a more reliable guide to progress than serum enzyme levels. Review of published reports suggests that overtreatment with corticosteroids may be a factor in chronicity of the disease and failure of adequate long-term response.

The childhood form of dermatomyositis is a well recognized entity characterized by muscle weakness and associated skin changes, and is usually classified with the collagen-vascular diseases. Although an autoimmune cause has not been proved, recent experimental studies on lymphocytes from patients (mainly adult) with polymyositis suggest the presence of a cell-mediated immunity (Currie *et al.*, 1971; Kakulas, Shute and Leclere, 1971; Johnson, Fink, and Ziff, 1972; Esiri, Maclennan, and Hazleman, 1973; Dawkins and Mastaglia, 1973; Haas and Arnason, 1974). The childhood form differs from the adult in a number of features, the most important perhaps being the lack of association with malignancy which occurs in about 20% of adult cases.

The disease has a natural tendency to remission and relapse, which makes assessment of new treatments difficult. However, earlier studies of its natural history highlight its high mortality as well as its tendency to chronicity, with associated contractures and persistent physical disability. Thus, Bitnum *et al.* (1964) quoted an overall mortality of one-third and an additional one-third with severe residual handicap, while Cook, Rosen, and Banker, (1963) had only 23 patients alive and well from a series of 50, whereas 15 had died and 12 were crippled. The introduction of corticosteroid therapy in the early 1950s was received with enthusiasm by some but others were more sceptical (Banker and Victor, 1966), and the mortality in several series published after its introduction remained relatively high despite the inclusion of treated cases: 11 out of 19 (Everett and Curtis, 1957), 10 out of 26 (Wedgwood, Cook and Cohen, 1953), 8 out of 31 (Muller, Winkelmann, and Brunsting, 1959) 2 out of 8 (Thieffry *et al.*, 1967), 2 out of 13 (Hill and Wood, 1970), 3 out of 22 (Roget *et al.*, 1971), 6 out of 22 (Ansell, Hamilton, and Bywaters, 1973).

Evaluation of corticosteroid therapy, and of other, subsequent immunosuppressive therapies, remains difficult for a number of reasons. Most personal series of cases are small, whereas the larger series published are usually retrospective, often poorly documented, and lacking details about the duration of the disease before treatment and objective quantification of disability before and after treatment. A number of fashions, however, seem to have evolved in relation to steroid therapy which are generally supported by most recent authors (Thieffry *et al.*, 1967; Stögmann, 1971; Vignos and Goldwyn, 1972; Sullivan *et al.*, 1972; Miller, 1973; Schaller, 1973; Haas, 1973; Benson and Aldo, 1973; Rose, 1974)—namely (1) high-dosage corticosteroid therapy is essential to produce remission; (2) initial high-dosage therapy needs to be continued until full remission is produced; (3) subsequent long-term low-dosage maintenance therapy is essential to prevent relapse; (4) response to therapy can be monitored by the fall in serum enzyme levels.

Thieffry *et al.* (1967), reporting 8 cases of dermatomyositis in childhood, concluded that corti-

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costeroids were the treatment of choice so long as the prednisone was given in high enough dosage of 2, 3, or even 4 mg/kg per day. Two of their 8 cases developed calcinosis. Sullivan *et al.* (1972) gave a detailed account of their personal experience of 18 cases of dermatomyositis in childhood. 12 of their 18 cases had an initial daily dosage of 2 mg prednisone/kg, 3 a dosage of 1·5 mg/kg, and 3 a dosage of 1 mg/kg. This was followed by a daily maintenance dose of 0·25 mg/kg. The maintenance dose stage of treatment was reached only after at least 12 months in all cases and the total duration of therapy was at least 2 years in all cases. 7 of the 18 cases developed calcinosis.

Of 16 cases of childhood polymyositis reviewed by Rose (1974), 10 had been given large doses of prednisone (60–80 mg/d) followed by at least 2 years of maintenance therapy (usually 10–15 mg/d), and 6, diagnosed before 1953, had received no corticosteroids and only supportive therapy. 7 of the 10 patients treated with corticosteroids who were treated within 1–6 months of onset of symptoms had a short period of severe disability; 6 made an excellent recovery and 1 died. 3 patients treated 17–36 months after onset made a less complete but functionally satisfactory recovery. The 6 untreated cases had a much longer period of severe disability but eventually made a good recovery. Calcinosis was present in 50% of both the treated and untreated patients. Rose concluded that 'the aim of treatment is not to eradicate all manifestations of the disease process, but to maintain patients in an ambulatory and independent functional state during the active phase of the disease,' and that 'the duration of the active phase of the disease has been found to be 2–3 years in most cases and therefore maintenance therapy is recommended for this period of time for prevention of relapses.'

My policy over the past 10 years has been to try to obtain a remission with a relatively moderate daily dose of prednisone of about 1–1·5 mg/kg rather than the 1·5–2 mg/kg or more used by most authors, and to get the patient completely off the steroid therapy as soon as possible by gradually tapering the dosage as soon as there is clinical *improvement* without waiting for full clinical remission. Reduction in steroid dosage has to be slow (usually 2·5 mg every fourth day), and any sign of deterioration is treated by reverting to the previous dose point. During this phase of treatment close clinical monitoring of the patient, preferably at weekly intervals, is an advantage and the dosage has to be carefully tailored to the response.

I think it unnecessary, and possibly less reliable, to monitor progress from the levels of serum enzymes rather than from the clinical response. In some children the level of creatine phosphokinase (and other enzymes) may remain normal throughout the acute phase. The erythrocyte sedimentation rate (ESR) may also be normal in the acute phase. Moreover, when enzyme levels are raised their fall may lag behind clinical remission. Similarly, while corticosteroid therapy is being reduced there may be a rebound in the serum enzyme levels, which most authors seem to regard as an indication for once again stepping up the steroid dosage. So long as clinical improvement is maintained I think it is reasonable to continue reducing the steroid dosage.

Early diagnosis and treatment are probably important in obtaining a good response and preventing some of the chronic changes in the muscle. Diagnosis may be difficult when serum enzyme levels and ESR are normal. The presentation may often be atypical, but the clinical picture of weakness, usually of recent onset, together with general malaise and misery should alert one to the possibility of dermatomyositis. An electromyogram (EMG) will usually show an abnormal myopathic pattern of change, often associated with spontaneous fibrillation potentials suggesting associated denervation. Muscle biopsy in some cases is unequivocally abnormal but in others it may be normal or show only focal inflammatory response or a perifascicular atrophy (Dubowitz and Brooke, 1973), or evidence of vasculitis (Banker and Victor, 1966; Banker, 1975). A therapeutic trial is worthwhile as soon as clinical diagnosis is made, and one should not depend on the confirmation by special investigations.

The present report is of a personal series of 8 consecutive cases of juvenile dermatomyositis treated with a moderate dosage, short-term corticosteroid regimen.

Case histories

The essential clinical features and therapeutic regimen of the 8 cases are summarized in Tables I and II. For economy of space only two selected case summaries are reported (see Appendix). Detailed case summaries of all 8 cases are available on request from the author.

In each case there were general symptoms of malaise and misery in addition to muscle weakness. In some the onset of muscle weakness was clearly defined, whereas in others it seemed to be insidious and more protracted.

All cases showed subjective as well as objective signs of improvement within about 3 weeks of starting therapy, after which the corticosteroid was gradually tapered off and stopped within 6 months (or less). The course of maintenance steroid treatment was relatively long in case 4 because of two episodes of laryngotracheobronchitis with tracheostomy and subsequent abduction

TABLE

Clinical features of 8 cases of

Case no.	Sex	Age at referral	Duration of symptoms	Referral diagnosis	Presenting			
					Muscle weakness	Muscle pain	Joint pain	Skin changes
1	M	6 y	6 w (? 1 y)	Miliary tuberculosis	+	−	−	+
2	M	2 y 10 m	? 1 y	Unable to walk	+	−	−	+
3	F	3 y 8 m	2 m	? Muscular dystrophy	+	+	+	+
4	F	3 y 6 m	21 m	? Myopathy	+	−	−	+
5	M	6 y	6 w	Muscular dystrophy	+	−	+	+
6	F	12 y	3 m	Depression	+	+	+	+
7	F	12 y	8 w	Dermato-myositis	+	+	−	+
8	M	8 y	18 m	Dermato-myositis	+	+	−	+

N, normal; +, present or abnormal; −, absent; ESR, erythrocyte sedimentation rate; CPK, creatine phosphokinase EMG, electromyogram.

TABLE

Therapeutic regimen in 8 cases of

Case no.	Weight (kg)	Prednisone initial dose		Onset of improvement	Duration of initial dose
		mg/24 h	mg/kg (approx)		
1	23	30	1·3	1 w	2+3 w
2	14	20	1·5	3 w	3 w
3	15	20	1·2	1 w	2 w
4	15	15	1·0	3 w	4 w
5	22	30	1·3	3 w	8 w
6	38	40	1·0	1 w	10 d
7	42	30	0·7	1 w	1 w
8	27	40	1·4	? 2 w	4 w

Symbols as in Table I.

paralysis of the cords, although there was no relapse in weakness of skeletal muscles at the time. The patient in case 8 had already been on long-term steroid therapy at the time of referral, but the clinical remission was improved by an initial increase and subsequent gradual tapering of the dosage. A number of recurrences of weakness in Case 1 were associated with upper respiratory infection or bouts of diarrhoea, but each resolved on a short course of prednisone for 2 to 3 days. Several episodes of weakness in Case 4 occurred in association

I

dermatomyositis in childhood

ymptoms and signs				Investigations			
Respiratory difficulty	Swallowing difficulty	Contractures	General symptoms	ESR	Serum CPK level	EMG	Muscle biopsy
+	−	−	+	up	up		+
−	+	−	+	N	up (SGOT)		
−	−	−	+	N	N		
+	−	−	+	N	N	N later +	N
−	−	+	+	up	up	+	+
+	−	+	+	N	N	+	N
−	−	−	—	up	up	+	
−	−	+	+	up	up	+	

II

dermatomyositis in childhood

Total duration of prednisone	Current status	Relapses	Duration of follow-up	Comments
6 m	Complete remission	+	13 y	Haemorrhagic chickenpox 2 weeks after treatment started; relapses associated with respiratory or intestinal infections; responded rapidly to 3 days' prednisone
4 m	Complete remission	−	7 y	Rapid resolution except for skin changes; no relapse off treatment
6 m	Complete remission	−	4 y	Rapid remission; skin rash took several months
21 m	Complete remission	+	3 y	2 attacks of acute laryngo-tracheobronchitis needing tracheostomy; prolonged abductor paralysis of cords; short relapses of weakness; responsive to steroids
6 m	Complete remission	−	2 y	Rapid resolution; no recurrence
14 w	Complete remission	−	1 y	Rapid resolution including contractures of elbows and depression (see Appendix)
5 m	Complete remission	−	3 y	Rapid remission; skin changes persisted several months
18 m	Almost full remission		2 y	Still on maintenance steroids; gradually reduced from 17·5 mg/d at referral to 4 mg/d at present (see Appendix)

with the tapering of the long-continued maintenance therapy. They usually responded rapidly to a one-step increment (usually 2·5 mg/day) in the prednisone dosage. The patient did not have any overt relapse of her dermatomyositis. None of the cases developed calcinosis.

Discussion

Although the series of cases is small it has the advantage of personal and uniform management. Most previous reports of personal series of cases

have averaged about one case a year over the period covered. This may reflect the true rarity of the condition or suggest that cases are being treated (or missed) by general practitioners or paediatricians and not referred to regional centres.

Our experience shows that a complete remission can be obtained in dermatomyositis in childhood with moderate doses of corticosteroid and that it is better to get the child off treatment as soon as possible. None of these patients has developed calcinosis. Possibly the protracted course of the disease reported by many authors may have resulted from the treatment rather than the disease itself and may have reflected a steroid-induced myopathy from high-dosage, long-term treatment. This is also reflected in some of the recent case reports advocating other forms of immunosuppressive therapy in cases of dermatomyositis that have ostensibly failed to respond to corticosteroids.

Thus one of the 4 cases of polymyositis treated with azathioprine by Benson and Aldo (1973) was a 10 year-old girl who presented with proximal weakness, periorbital oedema and discolouration, and a confluent rash of the limbs and buttocks. The duration of the illness is not given. High-dosage prednisone (2 mg/kg) for 3 months did not produce remission but caused pronounced hypertension, Cushing's syndrome, and epistaxis. Azathioprine was then given for 24 months, during which the rash resolved and the muscle power steadily improved. Details are not given in the text, but judging from the chart of the clinical course prednisone was continued at 40 mg/day for a further 2 months, then at 80 mg on alternate days for 2 months, followed by 40 mg daily for 3 months, 80 mg alternate days for a further 1½ months, and then very gradually tapered over 20 months. The power seems to have reached a normal level after about 18 months from the start of treatment.

Haas's (1973) series of 8 cases treated by various courses of immunosuppressive drugs included 2 children. One (Case 2), an 8-year-old boy, had presented at the age of 17 months with generalized weakness. Three muscle biopsies were unremarkable, the SGOT level was slightly raised, and an EMG suggested myopathy. Within two months he was unable to stand and needed tracheostomy. Gradual improvement followed prednisone therapy, which was continued for 8 months and subsequently restarted after an exacerbation soon after stopping, and then continued for 4½ years despite excellent remission. 2 months later a moderate exacerbation was treated with azathioprine. The other child (Case 7), a 12-year-old girl, had presented with weakness, aching legs, and a transient rash. Because a month of high-dosage prednisone therapy (up to 60 mg daily) failed to lower the serum enzyme levels azathioprine was added and continued for 5 months. The enzyme levels decreased but remained above normal and power did not change. A 6-week course of intermittent (once or twice weekly) methotrexate was added. Over the next 6 months she slowly deteriorated and became chairbound. Cyclophosphamide with prednisolone was then given. There was some drop in CPK level but she remained unable to walk.

In a retrospective 'review of the charts' of 23 children with dermatomyositis attending the Children's Hospital at Stanford between 1958 and 1973, Miller (1973) tried to assess the value of corticosteroid therapy. The group was a heterogeneous one without uniformity of management or follow-up. 8 were referred before treatment as diagnostic problems, 10 were referred after treatment had begun, and 5 had already been treated for more than a year. Out of 14 with 5 or more years' duration one had died, 11 of the remaining 13 had residual physical damage interfering with function to some degree, while in 4 the disease had progressed. It is impossible from the data given in this paper to draw any correlations between the clinical status and therapeutic regimen in individual cases.

Conclusions

Dermatomyositis in childhood is a corticosteroid-responsive disease. Treatment should be started as soon as a clinical diagnosis is made, even if the sedimentation rate and serum enzyme levels are normal and a muscle biopsy negative. Prednisone in moderate dosage (1–1·5 mg/kg) given daily in divided doses, is the treatment of choice. The dose is an arbitrary one and may be rounded up or down to the nearest 5 mg for ease of dispensing. At the first signs of improvement in muscle power, usually accompanied also by some improvement in the patient's general well-being, the dosage should be steadily tapered at a rate of, say, 2·5 mg every fourth day. If there is any suggestion of regression in symptoms the dose should be stepped up by 2·5 mg and retained for a further week before again reducing it. Prednisone may be stopped completely once it is down to 2·5 mg/day and remain stopped if improvement is maintained.

If there is no response at all to prednisone within say 4–6 weeks a more prolonged course is unlikely to be beneficial and it should be gradually tapered down as before and alternative forms of treatment (azathioprine, cyclophosphamide, methotrexate) tried. The diagnosis should also be reviewed and

alternatives such as viral myositis (due for example to Coxsackie B4) excluded. Indeed, testing for antibody levels to various potential viral agents causing myositis is worth while in all cases of apparently classical dermatomyositis at presentation.

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Correspondence to Professor V. Dubowitz, Department of Paediatrics and Neonatal Medicine, Hammersmith Hospital, Du Cane Road, London W12 0HS.

Appendix

Case histories.

Case 6. This 12-year-old schoolgirl presented with a 4-month history of depression, shortness of breath on exertion, gradual loss of weight, aching of the shoulders when carrying a schoolbag, and a change in her gait with walking on her toes. She also noted aching in the thighs and calves at times and an inability to straighten her elbows fully, with some associated pain in the elbow joint. Any activity beyond her routine led to rapid exhaustion. When examined she tended to walk on the toes and could not walk on her heels. She could go up and down stairs but refused to get up from the floor. The elbows showed about 10° limitation of extension and some pain when trying to extend them. There was also some limitation of flexion of the spine. There was no joint swelling or tenderness. The muscles were not tender to palpation. The tendon reflexes were all present. A band of violaceous colour was present along the lower half of both eyelids, and there was also slight erythema of the malar region of the face. On the basis of her general malaise in association with a suggestion of muscle weakness and the skin change dermatomyositis was diagnosed.

The sedimentation rate was 10 mm in 1 hr and the serum CPK 40 IU/l. EMG showed fibrillation at rest and a myopathic pattern of small polyphasic potentials on volition. Biopsy of the gastrocnemius muscle two days after treatment was started seemed histologically and histochemically normal. Treatment was started immediately with prednisone 10 mg four times daily (1 mg/kg per day). Improvement started within 2 days, and when reassessed a week later she was walking better, found it easier to dress herself, had an improved range of extension of the elbows, was able to go up and down steps without difficulty, and was able to get up from the floor, although still with some difficulty. In addition, she was a changed personality—bright, cheerful, and co-operative—and had felt well enough to enter for her school examinations, from which she had previously had to withdraw because of her illness.

The prednisone was reduced by two decrements of 5 mg at 4-day intervals to 10 mg three times daily, after a further 2 weeks by 5-mg decrements at weekly intervals to 5 mg three times daily, and then by 2·5 mg decrements at weekly intervals until it was completely stopped about 4 months after starting therapy. She was completely well and was participating in various sports and coping well with both intellectual as well as physical activities. There has been no recurrence of symptoms over the ensuing 3 years.

Case 8. This 8-year-old boy was seen by a paediatrician in February 1973 with a history of discolouration around the eyes for about 8 months, followed by general depression, aches in the legs and difficulty with walking and going up steps for 2–3 months, and discolouration over the knuckles, knees, and elbows for about 3 weeks. On examination at that time there was skin discolouration and muscle weakness and enlargement of the liver and spleen.

The ESR was 45 mm in 1 hr and the CPK was raised to 140 IU/l.

Prednisone was prescribed at 40 mg/day. Four weeks later he was much improved, was able to dress himself and to trot, but not run. His temperament was improved and he tired less. The prednisone was rapidly cut to 20 mg/day and then to 15 mg and 12·5 mg after two further intervals of 3 weeks and continued at that dose. 2 months later, in early June, he had difficulty in getting up from the supine and in going up stairs. Prednisone 12·5 mg/day was continued. By the end of June progress was less satisfactory, and he was given methotrexate 3 mg/week for 11 weeks with no benefit. At the end of August prednisone was raised to 17·5 mg/day. This resulted in definite improvement and he was kept at that dose for 2 months before gradual reduction to 13 mg/day. When first seen at Hammersmith in mid-November 1973 he still had the violaceous discolouration of upper eyelids and erythema over elbows, knees, and knuckles. There was proximal weakness (MRC grade 3–4) of shoulders and hip-girdle muscles and some limitation of supination of the right forearm. He was feeling well in himself. EMG of the deltoid and quadriceps showed unequivocal myopathic changes. The CPK was normal.

In view of the apparent continued activity of the dermatomyositis it was decided to increase the prednisone to 20 mg/day with a view to improving his remission and then trying to taper it further down than before. Two weeks later he was much improved but not completely well. Prednisone was continued for 4 weeks at the same dose with marked improvement and almost complete resolution of weakness, and also some fading of the skin changes. Prednisone was then reduced by 2·5 mg decrements at 2-week intervals with no problem until down to 7·5 mg/day, when he became less active. The dose was raised to 10 mg/day with some improvement. When reduced to 7·5 mg he again became slightly weaker and it was raised again to 10 mg. In December 1974 he was still getting up from the floor and going up and down steps with only minimal difficulty. His skin lesions were still florid. The dose was reduced to 7·5 mg/day with no increase in weakness, but when reduced further to 5 mg/day he became weaker and the rash more florid. The dose was again increased to 7·5 mg/day and subsequently tapered very slowly by 1 mg every 2 weeks. He is currently down to 4 mg/day and remains fully mobile.

This child's dermatomyositis appears to have been fully responsive to prednisone initally and his subsequent relapse and difficulty with management may relate to the prolonged maintenance at 12·5 mg/day. Increasing the dose has produced further regression of symptoms and allowed a gradual tapering of the dose thereafter.