Sir,

This is in reference to the article ‘Therapeutic trials for systemic sclerosis: An update’ by Sardana and Garg.

The comprehensive review needs the following references [2-8] to be added.

It may also be added that as many as 14 dermatologists in various institutions have used dexamethasone-cyclophosphamide pulse therapy (DCP)/dexamethasone pulse therapy (DP) regimen for about 300 systemic sclerosis patients and found satisfactory recoveries. DCP therapy consists of transfusing 100 mg dexamethasone dissolved in 500 ml 5% glucose over one to two hours, for three consecutive days every month, along with cyclophosphamide 500 mg on day one in the same drip and 50 mg daily, orally, in between the pulses. In DP only dexamethasone 100 mg is transfused in 500 ml 5% glucose over one to two hours for three consecutive days every month. It is used in those patients where cyclophosphamide is contraindicated.

For the information of the dermatologists, DCP / DC is able to bring about a total/almost total reversal of the changes, including skin hardening, pigmentary changes, arthritis, dysphagia, dyspnea to a variable extent, gangrene, fingertip ulceration, and even Raynaud’s phenomenon, which are commonly observed in progressive systemic sclerosis patients. The total treatment is to be given for three to four years. Less doses as used by those who have found unsatisfactory results, are obviously due to inadequate treatment.

Ramji Gupta

President, Pemphigus and Pulse Therapy Foundation, New Delhi, India

Address for correspondence: Dr. Ramji Gupta, M-54, Jal Vihar Road, Lajpat Nagar-II, New Delhi - 110054, India.

E-mail: drramjigupta@yahoo.co.in

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Authors’ reply

Sir,

We are thankful to the author [1-2] for evincing interest in our article.

1. Firstly we focused on newer approaches in scleroderma. Dexamethasone-cyclophosphamide pulse (DCP) therapy is by no means a new approach.

2. The evidence against use of steroids is overwhelming. There is a mountain of evidence from textbooks, and guidelines of medicine, rheumatology and dermatology detailing the evidence against the use of steroids except for alveolitis, myocarditis and sometimes for renal involvement [14-22] (Table 1).

3. Steroids have multitude of side effects which add to the already multisystem damage of scleroderma. [13-15]

4. Skin improvement, which is a tool observed by most Indian case reports, is the most nonspecific tool to monitor improvement. Steroids per se have no role to play in altering the skin pathology of progressive systemic sclerosis (PSS). [14] Glucocorticoids are not effective in improving or preventing skin induration and the progression of systemic sclerosis (SSc; also known as scleroderma). [14]

5. The most crucial aspects is that evidence-based
Lastly, the author has excluded two articles reporting the side-effects, which have been reported from India.\[20,21\] This highlights the risks involved in the indiscriminate use of this form of therapy.

In view of the huge data from scientific journals, specialty books and international guidelines,\[3-17,21\] the obstinate persistence of DCP pulse in scleroderma is purely a individual perception which is beyond scientific purview as its role has not been mentioned in any evidence-based data to date.

The summary guidelines on steroids gleaned from the wealth of data are given below [Table 2].

### Table 1: Role of steroids in scleroderma

<table>
<thead>
<tr>
<th>Scientific data</th>
<th>Use</th>
<th>Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scleroderma Foundation</td>
<td>No use</td>
<td>Can be used for myocarditis, alveolitis</td>
</tr>
<tr>
<td>Evidence Based Dermatology</td>
<td>No use</td>
<td>Can be used for myocarditis, alveolitis</td>
</tr>
<tr>
<td>Cochrane Database</td>
<td>Not disease modifying</td>
<td>-</td>
</tr>
<tr>
<td>BAAD Guidelines</td>
<td>Not disease modifying</td>
<td>Can cause scleroderma renal crisis</td>
</tr>
<tr>
<td>ACR Guidelines</td>
<td>Not disease modifying</td>
<td>Can be used for myocarditis, alveolitis</td>
</tr>
<tr>
<td>American Academy Guidelines</td>
<td>Not disease modifying</td>
<td>-</td>
</tr>
<tr>
<td>European Academy Guidelines</td>
<td>Not disease modifying</td>
<td>Gives a sense of well being</td>
</tr>
<tr>
<td>Kelley’s Textbook of Internal Medicine[7]</td>
<td>Not disease modifying</td>
<td>-</td>
</tr>
<tr>
<td>Scleroderma: In Samter’s immunologic diseases[8]</td>
<td>Not disease modifying</td>
<td>-</td>
</tr>
<tr>
<td>ACP Medicine[6]</td>
<td>Not disease modifying</td>
<td>Can be used for myocarditis, alveolitis</td>
</tr>
<tr>
<td>Harrison’s principles of internal medicine[12]</td>
<td>Not disease modifying</td>
<td>Should not be given</td>
</tr>
<tr>
<td>Rheum Dis Clin North Am[13-16]</td>
<td>Not disease modifying</td>
<td>Should not be given</td>
</tr>
<tr>
<td>Oxford Textbook of Rheumatology[20]</td>
<td>Not disease modifying</td>
<td>Should not be given</td>
</tr>
</tbody>
</table>

### Table 2: Summary of dose effect and relationship of steroids\[1-8,12-16,20\]

1. Low-dose prednisone (10 mg/day or less)—edematous phase (skin involvement); joint and tendon pain.
2. High-dose prednisone (20-30 mg/day) with steroid-sparing agent such as methotrexate or azathioprine—inflammatory myositis, pericarditis, early active alveolitis.
3. Glucocorticoids have been associated with the development of renal crisis.\[8,6\]
4. Diffuse cutaneous SSc showed a significant association between prior high-dose glucocorticoids (prednisone 15 mg/d) and the development of scleroderma renal crisis.

Also, all our references were of evidence-based double blinded trials, whereas the references alluded by the author\[4-10\] are not.

Secondly, the indexed literature does not contain references 4, 6, and 8, referred to by the author.

And case reports are not in any way considered as evidence even in the Cochrane registry of controlled trials, and most of the data reported by the author do not meet the standards of the Cochrane guidelines.

Lastly, the author has excluded two articles reporting the side-effects, which have been reported from India.\[20,21\] This highlights the risks involved in the indiscriminate use of this form of therapy.

In view of the huge data from scientific journals, specialty books and international guidelines,\[3-17,21\] the obstinate persistence of DCP pulse in scleroderma is purely a individual perception which is beyond scientific purview as its role has not been mentioned in any evidence-based data to date.

The summary guidelines on steroids gleaned from the wealth of data are given below [Table 2].

Kabir Sardana, V. K. Garg
Department of Dermatology and STD, Maulana Azad Medical College and Lok Nayak Hospital, Delhi, India
Address for correspondence: Dr. Kabir Sardana, 466, Sector 28, Noida, UP - 201 303, India. E-mail: kabirijdvl@gmail.com
REFERENCES


