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**Case Report**

# Managing the challenging cases of pemphigus vulgaris with modified pulse therapy: case series

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## ABSTRACT

In pemphigus vulgaris, pulse therapy is a promising modality to achieve better therapeutic response and to minimize the side effects of daily steroids. Here we report 5 different challenging situations where we successfully managed with modification of pulse therapy. Our first case had multiple comorbidities with persistent skin infections managed with dexamethasone and cyclophosphamide pulse (DCP) and interval pulse. The second case was unmarried hypertensive male with poor DLQI managed with Azathioprine pulse therapy. The third case was uncontrolled diabetic patient with pulmonary tuberculosis managed with rituximab. The fourth case was diabetic and hypertensive was managed with one day dexamethasone cyclophosphamide pulse therapy (DCP). The fifth case was not completed family with cushingoid features managed with dexamethasone azathioprine pulse therapy (DAP). Pulse therapy is effective in controlling disease activity in pemphigus. But it can be modified in some situations according to the patient needs.

**Keywords:** Pemphigus, Modified pulse therapy, DCP

## INTRODUCTION

Pulse therapy is defined as administration of suprapharmacological dosage of drugs in an intermittent manner.[1] Dexamethasone cyclophosphamide pulse (DCP) was first introduced by Pasricha and Gupta in pemphigus vulgaris patients. Dexamethasone 100 mg in 500 ml of 5% glucose as a slow intravenous infusion over 2 hours on 3 consecutive days. On the second day, cyclophosphamide 500 mg is added to the infusion. This constitutes one cycle of DCP. One such cycle of DCPs are repeated at every 28 day intervals counted from the first day of the pulse therapy. In between the pulse therapy, the patient receives oral cyclophosphamide 50mg daily. The DCP regimen constitutes of four phases. In phase I, the patient may develop new lesions in between the DCPs and can therefore be given additional treatment (conventional daily oral corticosteroids or additional dexamethasone interval pulses) to fasten the recovery.[2] The patient is considered to have entered phase II, when the skin and the mucosal lesions have healed completely. In phase 2, the patient does not have any new clinical lesions but receives 9 such cycles of DCPs along with cyclophosphamide 50 mg orally per day. In phase III, the patient receives only cyclophosphamide 50 mg orally per day for 9 months. In phase 4, patient is followed up for next 10 years to look for any clinical relapse.

## CASE REPORT

### *Case 1*

A 50 year old post menopausal female had co-morbidities such as diabetes, hypothyroidism, anemia, hypoproteinemia, hyponatremia with persistent

pseudomonas and MRSA skin infections. She was managed with Injection Piperacillin and Tazobactam 4.5 g iv thrice daily for 10 days followed by injection Meropenem 500 mg iv thrice daily for 10 days, correction of fluid and electrolytes imbalance, iron and folic acid supplementation, one unit of blood transfusion, 25% human albumin infusion given for 3 consecutive days and regular monitoring of blood glucose level following insulin therapy. Once the infection was controlled and other parameters were corrected patient was started on DCP therapy. Since patient was having persistent new lesions even after starting DCP, interval pulse was given. Following which, second pulse was given, patient started responding well.

***Case 2***

A 27 year old unmarried obese male found to be hypertensive with poor Dermatological Life Quality Index. First he was started on dexamethasone azathioprine pulse therapy following which he developed uncontrolled hypertension. Physician opinion obtained and ultrasonogram abdomen was done to rule out other medical causes of hypertension. His blood pressure was brought under control with anti-hypertensive medications. Then he was planned for Rituximab. But he developed hypersensitivity reaction following the first cycle of Rituximab infusion. So he was started on Azathioprine pulse therapy (Azathioprine 300 mg once in 28 days followed by daily Azathioprine 50 mg). Following 2 cycles of Azathioprine pulse therapy patient responded well.

***Case 3***

A 55 year old male had pulmonary tuberculosis, uncontrolled diabetes with urine ketones positive. He was managed with insulin therapy with strict blood glucose level monitoring was done. Patient was started on Anti tuberculosis therapy. During the intensive phase of ATT, he was started on daily steroids to control the disease activity. After completing the intensive phase of ATT, he was started on Dexamethasone cyclophosphamide pulse therapy. Since therapeutic response was not adequate following three cycles of DCP, he was started on Rituximab after obtaining chest physician opinion. After two doses of Rituximab patient had good therapeutic response.

***Case 4***

A 59 year old male with type 2 diabetes and hypertension on anti-diabetic and anti-hypertensive drugs. His blood pressure tends to shoot up on the first day of pulse therapy. So regular three days DCP therapy was modified to one day DCP therapy for this patient. Following five such cycles of one day DCP with daily oral cyclophosphamide 50 mg patient responded well with significant reduction in ABSIS was achieved.

***Case 5***

A 34 year old male, not completed family was on daily oral steroids for 1 year with cushingoid features. So initially he was started on azathioprine pulse therapy (Azathioprine 300 mg once in 28 days followed by daily Azathioprine 50 mg) however he did not have adequate clinical response for 2 months. So he was switched over to dexamethasone azathioprine pulse therapy. After 2 months of DAP therapy he was shown good clinical response.

## DISCUSSION

Pemphigus is a group of chronic autoimmune blistering disease characterized by the presence of antibodies against desmosome adhesion proteins. Dexamethasone cyclophosphamide pulse therapy have a potential to offer cure in pemphigus patients.[2] The advantages of DCP therapy are faster healing of lesions, faster control of disease activity, long term clinical recovery and avoidance of daily steroid side effects.[3]

Dexamethasone pulse causes shift of Th profile towards the Th2 profile decreases CD3, CD4 T-cell phenotypes. This recovers in 7 days without significant effect on B cells.[1,4] Intravenous cyclophosphamide causes maximum suppression of B-cells and moderate suppression of CD3 T-cells and natural killer cells. It takes two to four months to recover.[5]

In our cases, all the patients had multiple co-morbidities in which the regular DCP pulse was not able to administer. In our first case persistent infections are one of the relative contraindications for steroids, hence after controlling the infections as well as hypoproteinemia, patient was given pulse steroids. Because hypoproteinemia per se is the risk factor for infections as well as decreased response to protein bound drugs. In our second case, patient was unmarried obese and hypertensive, so he was started on Azathioprine pulse therapy. In our third case, patient was having pulmonary tuberculosis as well as diabetic so he was started on Rituximab. In our fourth case, patient was both diabetic and hypertensive, steroids have role in increasing both blood sugar levels and blood pressure so he was given only one day DCP followed by daily oral cyclophosphamide. In our fifth case, patient was not completed family so DAP therapy was given.

A review by Kaul et al have reported various modifications in therapy against pemphigus patients with co-morbidities.[6] A six year study by Rao et al also reported the modifications to the original DCP therapy protocol to be very effective and useful.[7] Our case reports also found that modification in pulse therapy to be effective against challenging situations in pemphigus vulgaris.

## CONCLUSION

Pulse therapy is effective in controlling disease activity in pemphigus. But it can be modified in some situations according to the patient needs. However further randomized trails are required to ensure the effectiveness of modification in pulse therapy against pemphigus patients with multiple co-morbidities.

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## REFERENCES

1. Pasricha JS. Pulse therapy in pemphigus and other diseases 2nd ed. New Delhi: Pulse Therapy and Pemphigus Foundation 2000.
2. Pasricha J S. Pulse therapy as a cure for autoimmune diseases. Indian J Dermatol Venereol Leprol 2003;69:323-8
3. Pasricha JS, Khaitan BK. Curative Treatment for Pemphigus. Arch Dermatol. 1996;132(12):1518–9.
4. Kricheli D, Mwaniki D, Zlotkin-Frušić M, Goldsmith D, Rabinov, et al. The distribution of pemphigus vulgaris-IgG subclasses and their reactivity with desmoglein 3 and 1 in pemphigus patients and their first-degree relatives. Br J Dermatol. 2000;143:337-42.
5. Reis VM, Toledo RP, Lopez A, Diaz LA, Martins JE. UVB-induced acantholysis in endemic Pemphigus foliaceus (Fogoselvagem) and Pemphigus vulgaris. J Am Acad Dermatol. 2000;42(4):571-6.
6. Kaul R, Sanjay CJ. Review on Pulse Therapy: A Novel Approach in the Treatment of Pemphigus Vulgaris. J Indian Acad Oral Med Radiol. 2010;22:211-4
7. Rao PN, Lakshmi TS. Pulse therapy and its modifications in pemphigus: A six year study. Indian J Dermatol Venereol Leprol. 2003;69:329-33.

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