

Letter to the Editor: High-Dose Intravenous Methylprednisolone for Pure Red Cell Aplasia

I have read with interest the case report of Needleman [1] "Durable Remission of Pure Red Cell Aplasia After Treatment With High-Dose Intravenous Gammaglobulin and Prednisone" in the recent issue of the *Journal*.

I have used high-dose intravenous methylprednisolone (HIVMP) (daily 30 mg/kg for 3 days, 20 mg/kg for 4 days, followed by 10, 5, 2 and 1 mg/kg per week for each, each dose given 2–5 minutes) for a 10 year old girl with pure red cell aplasia longer than 8 months duration, since it was used in the treatment of Diamond-Blackfan syndrome [2]. Her pure red cell aplasia was found unresponsive to conventional prednisone (2 mg/kg) for 6 months duration previously.

Normoblastemia (2%) with reticulocytosis (3.2%) was observed on the 12th day of HIVMP treatment. Hemoglobin increased to 11.34 g/dl (Hct:34%) on the 23rd day and reached 16.4 g/dl (Hct:50%) in 3 months with normoblastic erythroid hyperplasia of the bone marrow.

Although spontaneous recovery could not be rejected

as in Needleman's case, timing of the response may strongly suggest that HIVMP was effective in this case. She has been off the treatment more than 3 years and still enjoys a hematologically and clinically normal life.

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2. Özsoylu S: High-dose intravenous corticosteroid treatment for patients with Diamond-Blackfan syndrome resistant or refractory to conventional treatment. *Am J Pediatr Hematol Oncol* 10:210–217, 1988.

Author's Reply

The letter by Özsoylu suggests yet another option in approaching the patient with pure red cell aplasia (PRCA) whose disease fails to respond to glucocorticoids. This patient exhibited durable remission after extremely high doses of glucocorticoids were given intravenously and tapered over 5 weeks. The approach to the steroid refractory patient with PRCA has been recently reviewed by Dessypris [1]. Cytotoxic agents, antithymocyte globulin, plasmapheresis, splenectomy, IV Ig, and cyclosporin have all exhibited some efficacy in this situation. To this available armamentarium, perhaps such "industrial dose" escalations of glucocorticoids should be added. This modality would be inexpensive compared to IV Ig and would obviate many of the toxicities of the cytotoxic agents.

I have three reservations about this regimen as it is described, however: patients who fail a 3 to 6 month trial may exhibit considerable Cushingism and other steroid

toxicity which will be exacerbated by this regimen. IV glucocorticoid administration adds to cost and labor and may be no more efficacious than oral treatment. The regimen as described will almost certainly produce adrenal suppression and require careful slow tapering beyond 1 mg/kg to avoid adrenal insufficiency.

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1. Dessypris EN: Pure Red Cell Aplasia. Baltimore: John Hopkins University Press, 1988, pp 98-117.