

---

# Efficacy of intravenous immunoglobulins in livedoid vasculopathy: Long-term follow-up of 11 patients

Babak Monshi, MD, Christian Posch, MD, Igor Vujic, MD, Alma Sesti, MD,  
Silke Sobotka, MD, and Klemens Rappersberger, MD  
Vienna, Austria

**Background:** Evidence for the efficacy of various therapies of livedoid vasculopathy (LV) is limited.

**Objective:** We sought to determine efficacy and tolerability of 2 g/kg of intravenous immunoglobulins (IVIG) every 4 weeks in patients with LV.

**Methods:** This was a long-term follow-up study of 11 patients with LV treated with 2 g/kg of IVIG assessing the clinical characteristics, disease course, and quality of life.

**Results:** The treatment regimen led to complete remission of ulcerations and pain in 17 of 29 disease episodes (59%) after 3 cycles and in 25 of 29 episodes (86%) after 6 cycles. Two disease episodes showed remission after 7 and 8 cycles, resulting in a total number of remissions of 27 (93%). Subscore analysis showed resolution of pain in 80% after 2 IVIG cycles. Disease severity and quality of life were significantly improved after 6 cycles. Median duration of remissions was 26.7 months after initial and 7.5 months after subsequent disease episodes.

**Limitations:** This was a retrospective study that did not include the comparison of IVIG efficacy and its impact on quality of life with treatment options.

**Conclusions:** In our patients with LV, high-dose IVIG led to fast and complete resolution of pain and ulcerations and to substantial improvement in quality of life. (J Am Acad Dermatol 2014;71:738-44.)

**Key words:** atrophie blanche; Dermatology Life Quality Index; intravenous immunoglobulins; livedo reticularis; livedoid vasculitis; livedoid vasculopathy; quality of life; thrombotic vasculopathy.

Livedoid vasculopathy (LV) is an orphan disease characterized by the triad of livedo reticularis, chronic recurrent painful ulcerations, and stellate white scars with hyperpigmented borders, ie, atrophie blanche.<sup>1-4</sup> The major histopathologic findings are thrombotic occlusions of median-sized arterial vessels in the dermis. Various deviations in fibrinolysis or coagulation were found in such patients and have led to the notion that LV is a primary thrombo-occlusive disorder. However, up to 50% of patients lack any evidence of associated coagulation disorders. Nevertheless,

#### Abbreviations used:

DLQI: Dermatology Life Quality Index  
IVIG: intravenous immunoglobulins  
LV: livedoid vasculopathy  
LVAS: livedoid vasculopathy activity/severity score

therapeutic recommendations mainly refer to antiplatelet, antithrombotic, anticoagulant, and fibrinolytic medications, which are often unsatisfactory.<sup>3,5-8</sup> In contrast, beneficial effects of intravenous

---

From the Department of Dermatology and Venereology, Rudolfstiftung Hospital, Teaching Hospital of Vienna Medical University.

Funding sources: None.

Conflicts of interest: None declared.

Accepted for publication May 15, 2014.

Reprint requests: Babak Monshi, MD, Department of Dermatology and Venereology, Rudolfstiftung Hospital,

---

Juchgasse 25, A-1030 Vienna, Austria. E-mail: [babak.monshi@wienkav.at](mailto:babak.monshi@wienkav.at).

Published online July 9, 2014.

0190-9622/\$36.00

© 2014 by the American Academy of Dermatology, Inc.

<http://dx.doi.org/10.1016/j.jaad.2014.05.039>

immunoglobulins (IVIG) have been demonstrated in several case reports and in 1 open trial with low-dose IVIG.<sup>9-14</sup> The therapeutic mechanisms of IVIG in LV are unknown, but it can be hypothesized that in addition to its anti-inflammatory properties it might exert anticoagulation effects through: (1) inhibition of thrombogenic effects of antiphospholipid antibodies, (2) inhibitory effects on platelet adhesion, and (3) modulation of endothelial function.<sup>15-19</sup>

Herein, we report a 10-year follow-up study of 11 patients with LV, who experienced a total of 29 disease episodes, all of which were treated with a high-dose regimen of IVIG.

## METHODS

### Demographic data, clinical and laboratory findings

This study was approved by our institutional review board. From 2002 until 2012, 11 consecutive patients with LV and primary and recurrent disease flares received IVIG at a dose of 2 g/kg body weight over 2 or 3 consecutive days every 4 weeks over 6 months. Each case was diagnosed on basis of clinical presentation (livedo reticularis, atrophie blanche, pain, ± ulceration) and by histopathology. Analyzed data were taken from the medical records and missing data were obtained by interviewing patients at follow-up visits or by telephone. Dermatologic findings were photodocumented at every visit.

We assessed demographic data, general medical history, disease duration before diagnosis, localization of lesions, trigger factors, seasonal worsening, previous and concomitant treatments, and results of baseline investigations at the time of diagnosis (Table I).

## Assessment of efficacy of IVIG therapy

To quantify patients' response to IVIG treatment we established a LV activity/severity score (LVAS), which was determined at each visit when IVIG was administered (Table II). Moreover, patients completed a Dermatology Life Quality Index (DLQI) questionnaire twice in their first disease episodes, before the first and after the sixth IVIG cycle. The primary objective was to evaluate the efficacy of IVIG treatment, assessing disease severity (LVAS) before the first and after the sixth cycle. Each disease episode was evaluated separately and the numbers of IVIG cycles needed to achieve remission were recorded. Remissions were defined as LVAS less than or equal to 2, absence of pain, and complete healing of ulcers. Newly arising pain and/or ulceration and LVAS greater than 2 were considered as relapses.

## Statistical analyses

All statistical computations were done using software (SPSS Statistics 21.0, IBM Corp, Armonk, NY). Metric data such as age are presented as mean ± SD or, in case of skewed data, using median (minimum, maximum). Nominal data are presented using absolute frequencies and percentages. Statistical analyses are based on 29 disease episodes. Significance of changes in LVAS and DLQI score from the first to the sixth IVIG application was assessed using paired *t* tests. A *P* value less than or equal to .05 was considered significant.

## RESULTS

### Demographic data, clinical and laboratory findings

The mean age at diagnosis was 34 ± 12 years and the median disease history before diagnosis was

### CAPSULE SUMMARY

- Therapy of livedoid vasculopathy is challenging.
- A total of 2 g/kg body weight intravenous immunoglobulin led to resolution of pain and ulcerations in the vast majority of disease episodes and to a significant improvement of quality of life in 11 patients.
- High-dose intravenous immunoglobulins may be an effective treatment for livedoid vasculopathy.

**Table I.** Baseline investigations of patients at the time of diagnosis

Clinical and imaging investigations	Cranial MRI, venous duplex ultrasound, ankle-brachial index
Seroimmunologic screening	Antinuclear antibodies and subsets, antineutrophil cytoplasmatic antibodies, C3, C4, CH50, circulating immune complexes, cryoglobulins, cold agglutinin, antistreptolysin O, rheumatoid factor
Thrombophilia screening	Homocysteine, protein C and S activity, activated protein C resistance, antithrombin III, factor V Leiden, anticardiolipin and β2 glycoprotein antibodies, lupus anticoagulant

MRI, Magnetic resonance imaging.

Download English Version:

<https://daneshyari.com/en/article/6071640>

Download Persian Version:

<https://daneshyari.com/article/6071640>

[Daneshyari.com](https://daneshyari.com)