Surgical Treatment of Congenital Lymphedema

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KEYWORDS

• Lymphedema • Magnetic resonance lymphography • Lymph nodes transplantation • Microsurgery • VEGF-c • Lymphangiogenesis

KEY POINTS

- The ideal treatment for lymphedema of the limbs must restore function and normal cosmetic appearance.
- Human lymph nodes express high levels of the lymphatic vessel growth factor VEGF-c, which is responsible for stimulating lymphangiogenesis in the treated limb.
- Primary lymphedema can be associated with other organ malformations and genetic disorders: cancer and diseases of the central nervous system, lungs, heart, kidneys, and other organs can accompany lymphedema of sudden onset. The most prevalent hereditary disorders associated with lymphedema are Milroy disease, Meige syndrome, lymphedema-distichiasis, and yellow nail syndrome.
- Patients with hypoplastic forms of lymphedema on MRL are the preferred candidates for autologous lymph nodes transplantation.
- When the lymphedema is not too advanced, complete or near complete recovery is possible.

INTRODUCTION

The ideal treatment for lymphedema of the limbs must restore function and normal cosmetic appearance. Physiotherapy (manual drainages, pressotherapy, compression, bandages) is the usual treatment for chronic lymphedema and is considered by many as the only treatment for long-term management. It is not a curative therapy, but helps to control the evolution of the disease. Chronic lymphedema is a progressive condition, characterized by a degenerative and inflammatory process resulting in diffuse, irreversible tissue fibrosis. Surgical treatment is an alternative method of controlling chronic lymphedema.[1]

Precise diagnosis of lymphedema has achieved major progress: lymphangiography with oil and lymphoscintigraphy were once very useful, but these examinations have important drawbacks (eg, infection, pulmonary embolism). Magnetic resonance lymphangiography (MRL) with T2-weighted imaging has greater sensitivity, and allows complete visualization of the lymphatic system without any injection.[2]

Free autologous lymph node transplantation in hypoplastic forms of lymphedema is a new approach for lymphatic reconstruction, a more anatomic strategy compared with the multiple lymphovenous anastomoses. Recent findings on the growth hormones produced by the lymph nodes permit further understanding of the efficacy of these procedures. Three lymph node flaps can be used, depending on the affected segment and available donor sites. These flaps are located on

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the inguinal, thoracic, or cervical area. The transplanted nodes pump the extracellular liquid responsible for lymphedema formation, and contain germinative cells that improve immune functions (**Figs. 1** and **2**).

Human lymph nodes express high levels of the lymphatic vessel growth factor vascular endothelial growth factor (VEGF)-c. This growth hormone is responsible for stimulating lymphangiogenesis in the treated limb. Saaristo and colleagues[3] compared the production of this growth factor by different tissues of the immune and hematopoietic systems. Lymph nodes expressed the highest levels of VEGF-c among the tissues tested (**Fig. 3**).

Each living node contains a plexus between the lymphatic and venous systems. The transplanted lymph nodes also probably work as a biologic lymphovenous anastomosis (**Fig. 4**).

CLINICAL PRESENTATION

Alterations of lymph drainage induce stasis of the lymph and progressive tissue changes with enlargement of the subcutaneous tissue and thickening of the skin.[4] Secondary infections, immune disorders, and cosmetic and psychosocial impairment can severely affect patients with lymphedema.

Diagnosis of lymphedema is done mainly by clinical assessment. A detailed history, clinical

Fig. 1. A 34-year-old woman suffering from lymphedema since puberty, resistant to intensive physiotherapy.

Fig. 2. Patient in **Fig. 1**, 1 year after free node transplantation at the right inguinal region.