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CLINICAL ARTICLE

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Lymphedema microsurgery improved outcomes of pediatric primary extremity lymphedema

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Abstract

Background: Primary lymphedema is an anomaly of the regional lymphatic system with long symptom duration or severe lymphatic obstruction. Few microsurgical treatments for primary lymphedema have been reported. This aim of this study was to investigate the outcomes of microsurgical treatments in pediatric primary lymphedema patients.

Methods: Between 2013 and 2017, pediatric primary lymphedema patients who underwent either lymphovenous anastomosis (LVA) or vascularized lymph node transfer (VLNT) were retrospectively reviewed. Cheng's Lymphedema Grading, Taiwan Lymphoscintigraphy Staging and indocyanine green lymphography were used to select the procedures. No compression garments were used postoperatively. Outcome measurements included circumferential difference, episodes of cellulitis, and Lymphedema-specific Quality of life questionnaire (LYMQoL).

Results: Nine patients with mean age of 9.2 years (range, 2–19 years) with 11 lower and two upper lymphedematous limbs underwent 11 VLNT and two LVA. All VLNT flaps survived. At a mean 38.4-months (range, 16–63 months) of follow-up, the mean circumferential difference in nine unilateral lymphedematous limbs was improved by 6.7 ± 9.9% (p = .066). Two patients with bilateral lower limb lymphedema had mean limb circumference improvements of 1.3 and 6.5 cm, respectively. In nine limbs with cellulitis preoperatively, episodes of cellulitis decreased by 2.67 times/year (p = .007). At a mean 22.3-months of follow-up (range, 13–24 months), the LYMQoL overall score in 6 patients older than 7 years was improved by 3.2 ± 1.1 points (p = .007).

Conclusions: Lymphedema microsurgery significantly improved the episodes of cellulitis and quality of life without utilizing compression garments in pediatric primary lymphedema patients.

1 | INTRODUCTION

Primary lymphedema that presented with extremity swelling, sensation of heaviness, and occasional cellulitis (Domaszewska-Szostek, Zaleska, & Olszewski, 2016; Olszewski, 2008; Zampell et al., 2012) is estimated with an incidence of 1.15/100,000 (Smeltzer, Stickler, & Schirger, 1985). The extermity lymphedema could be classified by the age as follows: *congenital lymphedema* (within 2 years after birth), *lymphedema praecox* (bewteen 2 and 35 years), and *lymphedema tarda*, greater than 35 years (Rockson & Rivera, 2008). The causes may be hereditary (Rockson, 2001) including Turner's syndrome (Ruibal Francisco, Sanchez Buron, Pinero Martinez, Bueno Lozano, & Reverte Blanc, 1997), Milroy's disease (Mellor et al., 2010) and trisomy 18 (Tanriverdi, Ertan, Hendrik, Remberger, & Schmidt, 2005). The variable

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