

A Multi-Institutional Study on the Efficacy and Safety of Wearing a Custom-Made Compression Elastic Garment for 6 Months for Klippel-Trenaunay Syndrome with Venous Malformation

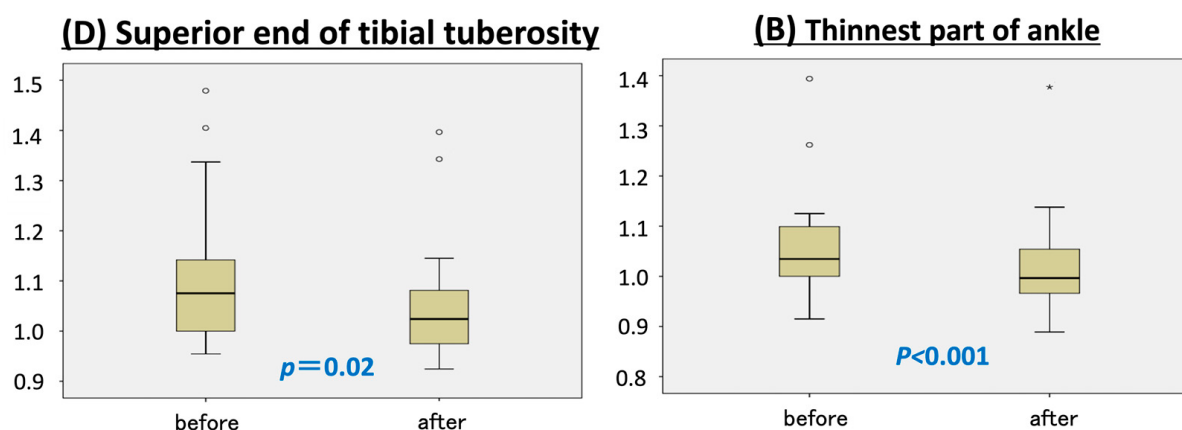
Miho Noguchi, Sadanori Akita, Fumio Nagai, Tadashi Nomura, Tsuyoshi Morishita and Shunsuke Yuzuriha (2025). Journal of clinical medicine

METHODS

- This multicenter, single arm, uncontrolled study was performed within four centers in Japan between 2021 and 2022. The main aim of this study was to assess the efficacy and safety of compression therapy using custom-made elastic CICATREX garments in patients with **Klippel-Trenaunay syndrome (KTS)**. KTS is a congenital vascular malformation syndrome involving low-flow vascular anomalies such as capillary malformations, venous and lymphatic malformations. Compression therapy has been reported to alleviate symptoms of low-flow vascular malformations in the extremities, including pain, swelling, ulceration, and coagulopathy.
- 20 patients (7 male, 13 female, mean age: 10.9 years) aged ≥ 2 years with KTS and venous malformation were included and were instructed to wear **CICATREX Airskin 30 mm Hg** garment (reaching at least the knee) during walking hours for 26 weeks (6 months).
- The **primary endpoint** was the **change in limb circumference ratio** (affected to unaffected limb) at four anatomical locations (B, B1, C, D) between baseline and 6 months.
- Secondary endpoints** included **change in pain** (1-5 scale), **patient's health condition** (modified Ranking Score (mRS)), **water content** (bioelectrical impedance), **garment elasticity** and **safety** (adverse events).

RESULTS

1 A significant decrease in limb circumference ratio



After 6 months, a **significant decrease in the limb circumference ratio** was observed at measurement points **B** (thinnest part of the ankle) ($p<0.001$) and **D** (superior end of the tibial tuberosity) ($p=0.02$). No significant changes in circumference ratio were observed at the remaining measurement points B1 (junction of the calcaneal tendon and gastrocnemius) and C (thickest part of the lower leg). These findings suggest that compression therapy may be more effective in thinner regions of the limb.

2 A slight improvement in pain and patient's health condition

Pain: After 6 months of treatment, pain while wearing the garment decreased from a baseline mean of 1.7 to 1.5, and pain without the garment declined from 1.5 to 1.44 on the 1-5 scale.

Patient's health condition: mRS is graded from 0 (no symptoms) to 5 (severe disability). The average mRS score improved from 1.6 to 1.4 after 6 months of treatment (1= no significant disability ; 2= slight disability).

3 No change in limb water content

No statistically significant differences in total or extracellular water content were observed between the baseline and post-treatment values in either the affected or healthy limbs.

4

A good tolerance

Overall, the use of the custom-made compression garments was well-tolerated and did not exacerbate any subjective symptoms. No severe adverse events related to garment use were recorded.

5

A decrease in garment elasticity

The elastic force of the garments significantly declined, measuring less than 50% of the initial force after 6 months of use ($p < 0.001$). The exact timing of elasticity loss remains uncertain and would require further study.

KEY MESSAGES

- ✓ Six months of wearing a custom-made CICATREX elastic garment significantly reduced limb circumference at two measurement points in patients with Klippel-Trenaunay syndrome and venous malformation.
- ✓ The garment was well tolerated.
- ✓ Given the observed decline in garment elasticity, replacement is recommended at least every six months to maintain therapeutic efficacy.
- ✓ Compression therapy using a custom-made elastic garment appears to be a safe and effective approach for managing limb overgrowth in patients with Klippel-Trenaunay syndrome and venous malformation. Future studies with larger cohorts are warranted.