Short-term application of complex decongestive therapy in a pediatric patient with Klippel-Trenaunay syndrome

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ABSTRACT

Klippel-Trenaunay syndrome (KTS) is a rare congenital disease characterized by vascular malformations and tissue hypertrophy. The recommended treatment to manage the symptoms of this syndrome is based on conservative methods including complex decongestive therapy. In this article, we report a pediatric case of KTS in whom short-term CDT was applied. A decrease in pain, fatigue, lymphedema, and an improvement in the functional mobility and lower extremity endurance were achieved.

Keywords: Congenital abnormality, conservative treatment, exercise, lymphedema, vascular malformation.

Klippel-Trenaunay syndrome (KTS) is a complicating disease characterized by lesions as port-wine stain which can be observed from birth. It may cause vascular and finger malformations, as well as bone and soft tissue hypertrophy. Although its etiology is still unclear, it is thought that it is manifested due to the mesodermal abnormalities which affect angiogenesis during the intrauterine development.

Port-wine stain is the first common finding detected in the distributed dermatomal areas in KTS. It is more prone to bleeding with increasing age, due to the capillary fragility and weakness in skin integrity. Abnormalities in platelets, coagulation, and fibrinolysis make the bleeding difficult to stop, and the affected area becomes vulnerable to recurrent skin infections.

Venous system disorders cause pain, lymphedema, functional limitations, and bleeding which are frequently seen in KTS. The most common symptoms are pain and lymphedema localized in extremities.

Elevation, compression, skin and wound care are the recommended primary treatment methods. Pulsed laser therapy, embolization, sclerotherapy, radiofrequency, and laser ablation can be also applied, if the symptoms progress. Open surgery consisting of vein stripping, stab phlebectomy, or more aggressive venous reconstructions are suitable for the patients who may not benefit from the minimal invasive or non-invasive techniques, such as complex decongestive therapy (CDT).

Complex decongestive therapy is a recommended conservative treatment method for KTS patients and consists of manual lymphatic drainage (MLD), skin care, compression, and lymphedema remedial exercises.

In this article, we report a pediatric case of KTS in whom short-term CDT was applied. Pain, fatigue and
lymphedema decreased meanwhile functional mobility and lower extremity endurance improved after the treatment.

**CASE REPORT**

An 11 and a half-year-old girl with a body weight of 35 kg, a height of 139 cm, a body mass index of 18.1 kg/m², and the right dominant side was admitted to the hospital with the rash in the lower extremities, high fever, dizziness, and recurrent cellulite infection for the last two years (three to four times a year). At the age of three, congenital edema, port-wine stains, diffuse-dilated varicose vascular structures, and hemangioma were observed in the subcutaneous adipose tissue in her bilateral lower extremities. However, the patient was not diagnosed until the age of five. The right/left vena saphena parva diameters were found to be 5 mm/6 mm in the Doppler ultrasound screening at the age of nine. Two years later, these diameters increased to 12 mm/8 mm. Lymphoscintigraphy findings were compatible with lymphatic obstruction in the bilateral lower extremities during her hospitalization. The patient was diagnosed with KTS by a specialist cardiovascular surgeon. Antibiotics, antithrombotic and venotonic drugs were started to prevent cellulitis and possible thrombosis and treat venous insufficiency. The patient was, then, directed to the physiotherapy for lymphedema. A written informed consent was obtained from each parent.

Physiotherapy assessments were performed by a trained lymphedema therapist. During the examination, syndactyly, port-wine stains, angiookeratomas, lymph fistulas, and cysts were inspected through different sides of her lower extremities. The dorsum of the left foot was more hypertrophic. Palpation of the lower extremities showed an increased skin temperature and the presence of non-fibrotic tissue. The Stemmer’s sign, which is a specific test of lymphedema, was positive. Congenital lymphedema was soft, localized at the knee level and below bilaterally, and triggered by infection with distal onset. During medical history taking, the patient described pain, burning, numbness, loss of strength, and fatigue spreading from the hip to the whole lower limb, particularly in prolonged standing and walking activities, with mobility limitation in daily life. The Wong-Baker FACES Pain Rating Scale (WBS), Visual Analog Scale-Fatigue (VAS-F), circumferential measurement (CM), Timed Up and Go (TUG) test, and 30-sec Sit-to-Stand test, which are all objective and reliable methods in children, were used to evaluate pain (in resting, activity and night), fatigue, lymphedema, functional mobility, and lower extremity endurance, respectively.

The patient received five sessions of CDT-Phase I consisting of MLD, compression bandage, skin care, and lymphedema remedial exercises. In each session, CDT was performed approximately 60 min. Briefly, a specific and individualized MLD scheme was applied according to the anastomosis ways, and a moisturizer containing water was applied for skin care before the bandaging. Inelastic compression bandages were used until above-knee bilaterally, according to the Földi approach. Consecutive active ankle, knee, and hip flexion/extension exercises were performed, and walking till pain limit was recommended. The patient and caregivers were instructed to perform the exercises for 20 reps for three times a day with a high-working pressure of compression bandages. In addition, home-based exercises and suggestions on daily life issues were given.

![Figure 1. (a) Before and (b) after the complex decongestive therapy.](image)

<table>
<thead>
<tr>
<th>Tests</th>
<th>Before treatment</th>
<th>After treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBS Rest</td>
<td>0/10</td>
<td>0/10</td>
</tr>
<tr>
<td>Activity</td>
<td>8/10</td>
<td>2/10</td>
</tr>
<tr>
<td>Night</td>
<td>0/10</td>
<td>0/10</td>
</tr>
<tr>
<td>VAS-F (cm)</td>
<td>9/10</td>
<td>2/10</td>
</tr>
<tr>
<td>TUG (seconds)</td>
<td>6.26</td>
<td>6.95</td>
</tr>
<tr>
<td>30-Seconds Sit to Stand test (repetitions)</td>
<td>14</td>
<td>16</td>
</tr>
</tbody>
</table>

WBS: Wong-Baker FACES Pain Rating Scale; VAS-F: Visual Analog Scale-Fatigue; TUG: Timed Up and Go Test.
Table 2. Lower extremity circumferential measurement values

<table>
<thead>
<tr>
<th>Circumferential measurement reference points</th>
<th>Right lower extremity (cm)</th>
<th>Left lower extremity (cm)</th>
<th>Difference (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metatarsophalangeal joint</td>
<td>21.6</td>
<td>21.8</td>
<td>-0.2</td>
</tr>
<tr>
<td>Ankle</td>
<td>27.4</td>
<td>26.8</td>
<td>0.6</td>
</tr>
<tr>
<td>4 cm</td>
<td>25.2</td>
<td>24.5</td>
<td>0.7</td>
</tr>
<tr>
<td>8 cm</td>
<td>26.2</td>
<td>25.1</td>
<td>1.1</td>
</tr>
<tr>
<td>12 cm</td>
<td>28</td>
<td>27.5</td>
<td>0.5</td>
</tr>
<tr>
<td>16 cm</td>
<td>29.2</td>
<td>28.6</td>
<td>0.6</td>
</tr>
<tr>
<td>20 cm</td>
<td>30.5</td>
<td>30.1</td>
<td>0.4</td>
</tr>
<tr>
<td>24 cm</td>
<td>30.5</td>
<td>31.1</td>
<td>-0.6</td>
</tr>
<tr>
<td>28 cm</td>
<td>31.4</td>
<td>31.5</td>
<td>-0.1</td>
</tr>
<tr>
<td>Total</td>
<td>250</td>
<td>247</td>
<td>3</td>
</tr>
</tbody>
</table>

At the end of CDT, a decrease in lymphedema, pain and fatigue severity, and an improvement in functional mobility and lower limb endurance were reported. The patient experienced some problems to afford the high-cost compression materials and could not achieve any lymphedema physiotherapist in her living place. Therefore, the patient’s parents had to undertake responsibilities about continuing Phase II of CDT as home-based physiotherapy program. It was planned to perform physiotherapy controls along with the medical examinations of the patient. The difference before and after treatment is shown in Figure 1. Pain, fatigue, functional mobility, and lower extremity endurance assessments are presented in Table 1 and the detailed CM results are listed in Table 2.

**DISCUSSION**

In this pediatric case of KTS, pain, fatigue and lymphedema decreased, and functional mobility and lower limb endurance improved after five sessions of CDT.

In the literature, it is reported that 38 to 93% of KTS patients face serious pain problems which are often localized at the lower extremity.[6,8,19,20] The KTS-related symptoms such as cellulitis, capillary malformation, chronic venous insufficiency, are shown to induce and increase the pain.[6,20] In this case, excessive pain was present before treatment due to vascular malformations and cellulitis, which is consistent with the literature.

Expanded and dysplastic veins due to KTS may cause fatigue, heaviness, pain, and lymphedema occurring in the lower extremity and significantly limit the activities of daily living of KTS patients, leading to functional disorders.[1,8,21] Patients have difficulty in walking long distances, climbing the stairs, and performing exercises, and the affected extremities get exhausted quickly.[6,21] Similarly, there was a decrease in the functional mobility as a result of the effects of pain and fatigue in this case. Therefore, TUG was used to evaluate the effectiveness of the applied CDT on functional mobility. According to the results, CDT improved the functional mobility, which makes it an effective method.

Previous studies have shown that CDT has pain-reducing effects in lymphedema treatment.[6,22] In addition to drug treatment, even in the short-term CDT application, the decrease in lower limb lymphedema may provide decreasing the pain during activity, and the heaviness causing fatigue during activities of daily living.

Manual lymphatic drainage is known as a pain-reducing treatment technique which enables the development of lymphangiomotoric and supports venous backflow.[22-24] It may have also provided pain relief in our case due to the stimulation of the parasympathetic system. However, current findings are still controversial whether MLD is effective in reducing lymphedema or not. Most of the studies are based on the fact that CDT is a holistic approach and compression therapy has been proposed to regulate the pain and lymphedema symptoms resulting from venous system problems; therefore, all parameters of CDT are important to gain better therapeutic results.[7,23]

Furthermore, exercise is an important parameter to treat muscle pump dysfunction in venous diseases by activating the lymphatic and venous
circulation. Since exercise is more beneficial and effective combined with compression, our patient and her caregivers were instructed to do it regularly on a daily basis, due to the chronic nature of KTS. In addition, we considered that regular exercise with compression bandage would be beneficial in decreasing lymphedema, pain, and fatigue for this case. Thus, the patient was referred to a lymphedema physiotherapist after discharge to complete the CDT.

This study is one of the rare studies in which CDT was used as a treatment method in pediatric KTS patient and the effects of short-term CDT treatment on pain, fatigue, lymphedema, functional mobility, and lower extremity endurance were examined. The limitation of the study was syndactyly which caused the insufficient finger compression application. Only short-term CDT treatment could be applied due to the lack of information about CDT and inability to achieve the physiotherapy. Therefore, this process may have reduced and resisted the effectiveness of CDT.

In conclusion, short-term CDT during hospitalization may improve pain, fatigue, lymphedema, functional mobility, and lower extremity endurance in pediatric KTS patients. Since KTS is a chronic condition, home-based suggestions and information should be emphasized. Although further large-scale and long-term studies using more objective evaluation tools are warranted, we suggest that both patients and physicians should be informed about CDT and CDT application is needed to be expanded.

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