

# Negative-pressure Wound Therapy for Sacral Pressure Ulcer in Gorham-Stout Disease

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**Summary:** Gorham-Stout disease (GSD) is a rare musculoskeletal disorder characterized by progressive bone resorption and overgrowth of lymphatic vessels. The mechanism of GSD is still largely unknown. Negative-pressure wound therapy (NPWT) is known to accelerate wound healing and is used worldwide. Herein, we report a successful treatment of a patient with GSD having a sacral pressure ulcer, using NPWT. An 18-year-old female GSD patient was referred to our department for treating a sacral wound. The wound was disinfected by pocket incision, cleansing, and administration of antibiotics; however, the lesion remain unhealed. Histopathology of the debrided sacral wound revealed fibrous granulation tissue, with no sign of lymphatic anomalies. NPWT was started with  $-75$  mm Hg of pressure, and neither lymphorrhea nor growth of lymphangioma was noted. Negative pressure was gradually increased to  $-125$  mm Hg. The ulcer size decreased to  $2 \times 2$  cm<sup>2</sup>, which healed 3 months after hospital discharge, with no recurrence for 8 months. For progressive diseases such as GSD, NPWT may cause the regrowth of lymphangioma or other neoplasms due to an increase in vessel endothelial growth factor. NPWT appears to be one of the safest and most effective wound therapies even for this rare and difficult disease, provided the use of the following treatment protocol: Pathohistological assessment before application of NPWT, and negative pressure initially set at a low level; then, gradually increased, with careful observation to avoid lymphorrhea. When changing the foam dressing, careful checking is important to determine whether the wound is necrotic, or if there is tumor-like tissue accumulation rather than healthy granulation. (*Plast Reconstr Surg Glob Open* 2021;9:e3303; doi: [10.1097/GOX.0000000000003303](https://doi.org/10.1097/GOX.0000000000003303); Published online 11 January 2021.)

Gorham-Stout disease (GSD), or simply Gorham disease, is a rare disorder of the musculoskeletal system, characterized by the proliferation of vascular channels that result in the destruction and resorption of osseous matrix. Since the first report by Gorham<sup>1</sup> over 50 years ago, little information about its mechanism and treatment has come to light. Patients with congenital lymphatic anomalies (including GSD) may experience problems with wound formation because osteolysis and lymphangioma may occur in the event of multiple lesions on the body.<sup>2</sup> Negative-pressure wound therapy (NPWT) is currently recognized as one of the most effective wound therapies.<sup>3</sup> We report on the successful application of NPWT for the treatment of a skin ulcer occurring on the sacral region in a patient with GSD.

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Received for publication June 30, 2020; accepted October 21, 2020.

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DOI: [10.1097/GOX.0000000000003303](https://doi.org/10.1097/GOX.0000000000003303)

## CASE REPORT

An 18-year-old female patient was referred for sacral swelling. The sacral wound was a pinhole-opened fistula surrounded by a 10-cm-diameter subcutaneous pocket (Fig. 1). Her body temperature was 38.3°C. She had been diagnosed with GSD during infancy at the National Children's Hospital in Tokyo, Japan, and since then her general condition has been monitored. She suffers from paraplegia and moves using a wheelchair. Blood test showed that her C-reactive protein concentration was 6.13 mg/dl, white blood cell count was 8100/μl, and hemoglobin level was 9.3 g/dl. Purple coloration of the skin was noted in her right hamstring area, suggesting the presence of venous hemangioma. As primary treatment, incision of the subcutaneous wound pocket, daily wound cleansing, and administration of cefazolin sodium 2 g/day were done, which improved her infectious symptoms. Wound bacterial culture revealed a small amount of *Staphylococcus epidermidis*. Computed tomography showed sharp caudal bone spines and surrounding bone resorption. No signs of a tumor-like lymphatic mass were noted. We suspected her wound to be a decubitus ulcer (due to pressure and friction around the deformed caudal spine) and conducted

**Disclosure:** The authors have no financial interest to declare in relation to the content of this article.

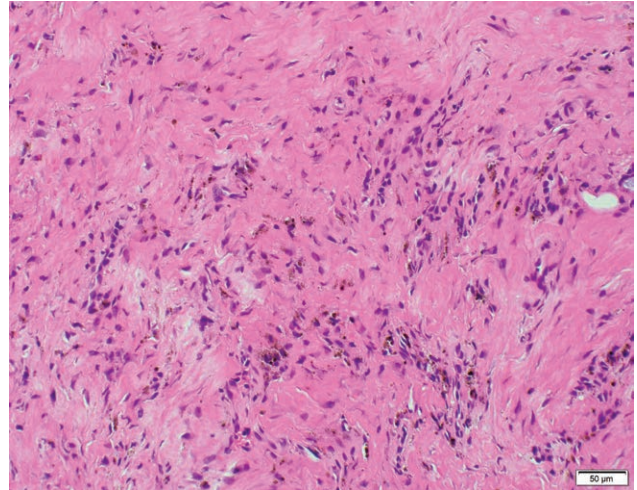


**Fig. 1.** An 18-year-old woman presented with a small fistula and surrounding subdermal pocket in the sacral skin. An yellow-to-purple area was noted on her right buttock, suggesting vascular abnormality.

debridement (including resection of the spine-shaped coccygeal bone, followed by pathological diagnosis of the wound) (Fig. 2). The histopathology of the excised specimen revealed fibrous granulation tissue, but no signs of tumor lesion (Fig. 3). These pathologic findings led us to apply NPWT to the wound lesion. The initial negative pressure was set at a low level of pressure (-75 mm Hg) to avoid the risk of hemorrhage or lymphorrhea. After confirming that there was no such leakage, the negative pressure was gradually increased to -125 mm Hg. The dressing of a polyurethane form was changed every 2-3 days, and no tumorous tissue was noted. The ulcer was granulated and reduced to a size of 2 × 2 cm<sup>2</sup> at day 18, at which point NPWT was terminated and the patient was discharged. The ulcer was eventually healed by ambulatory conventional



**Fig. 2.** Peri-operative image showing debridement for removal of bone protrusions.



**Fig. 3.** Microscopic view of the histopathological specimen (hematoxylin and eosin: ×200) showing granulative tissue without neoplastic changes, such as hemangioma or lymphangioma.

therapy at 3 months after patient discharge, with no recurrence over the 8-month follow-up period (Fig. 4).

### DISCUSSION

GSD, also known as vanishing bone disease or massive osteolysis, is a rare bone disorder characterized by progressive bone loss and the overgrowth of lymphatic vessels.<sup>1</sup> The exact cause of GSD is unknown, and no environmental or genetic risk factors have been identified.<sup>2</sup> Recent studies of the disease have shown increased serum levels of vessel endothelial growth factor C and interleukin 6 in GSD patients; however, it is unclear why local levels of these factors trigger abnormal angiogenesis and lymphangiogenesis at GSD lesions.<sup>2,4,5</sup> Therefore, a variety of treatments have been used and described by several researchers, including surgery, radiotherapy, and medications, alone or in combination, although an evident optimal treatment has not yet been established for GSD.<sup>2,6</sup> Systemic osteolysis and surrounding infiltrative soft-tissue lesions present risks of developing lymphedema, lymphorrhea, and, in



**Fig. 4.** Three-months postoperative image of the patient, showing wound healing.

more severe cases, chronic ulceration and angiosarcoma.<sup>2</sup> Thus, careful examination is necessary when treating a wound of a patient with GSD.

NPWT has been widely applied for wound treatment, especially for pressure ulcers<sup>3</sup> and diabetic foot ulcers,<sup>7</sup> and it is thought to have 4 principal mechanisms of action, as described in the literature: (1) contraction of the wound, (2) stabilization of the wound environment, (3) removal of extracellular fluids, and (4) micro-deformation at the foam–wound interface.<sup>3,7,8</sup> Because NPWT is known to increase interleukin 8 and vessel endothelial growth factor levels in wound edge cells,<sup>3,9</sup> NPWT could possibly cause lymphangioma growth due to increased vessel endothelial growth factor levels in patients with GSD. The levels at which these cytokines or growth factors cause tumors in GSD are unknown. In our case, NPWT application for a mean duration of 19 days resulted in no recurrence or proliferation of lymphangioma.

Katz et al study reported successful NPWT treatments after resection of lymphangiomas in children, though the negative pressure levels used therein were not mentioned.<sup>10</sup> For successful application of NPWT to such complex cases, we propose the following treatment protocol:

- 1) Pathohistological assessment should be conducted before choosing the application of NPWT, obtaining specimens such as a biopsy on initial debridement, to avoid risks of deterioration of the tumor lesion.
- 2) Gradually increasing the negative pressure level under careful observation for hemorrhaging or lymphorrhea, pain, and dermal complications.
- 3) When changing the foam dressing, special attention should be paid to determine if the wound is necrotic, or there is tumor-like tissue accumulation rather than a formation of healthy granulation.

### CONCLUSIONS

To our knowledge, this is the first study to report a successful NPWT treatment of a sacral wound in a patient

with GSD. With careful observation and management (including evaluation for lymphorrhea and lymphangioma recurrence), NPWT can be considered an effective and safe wound-treatment modality in patients with GSD, after pathological evaluation so as to avoid deterioration of the lesion.

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