Hay-Wells Syndrome is a rare genetic disorder characterized by ankyloblepharon, ectodermal dysplasia, and cleft palate. Recalcitrant scalp wounds with secondary infections are common. This case series describes the use of acoustic pressure wound therapy in 3-year-old fraternal twins (male and female) with HWS-associated scalp wounds. Present since infancy, the wounds were severe and extensive at presentation to the authors' wound clinic. Previous management consisted of standard topical treatments, including foam; oxidized, regenerated-cellulose/collagen with silver; calcium alginate; silver sulfadiazine cream; and biologic tissue matrix. Following admission to the authors' wound clinic, acoustic pressure wound therapy was administered one to three times weekly for 3 to 10 minutes for 7 months in addition to standard topical treatments to provide nonsurgical debridement and reduce wound bioburden without inflicting additional pain. Substantial improvements occurred during the first 5 weeks of consistent treatment. When treatments became sporadic due to health and family issues, wound deterioration occurred. After 7 months, wound sizes decreased by 31.3% in the boy and 1.1% in the girl, 70% of the wound surface in both children was covered with granulation tissue, and no clinical signs of infection were evident. The treatments were well tolerated. So far, the twins each received a total of 37 treatments. Consistent, long-term acoustic pressure wound therapy improved the status of severe, recalcitrant, Hay-Wells Syndrome-associated scalp wounds.

KEYWORDS: acoustic pressure wound therapy, AEC Syndrome, ectodermal dysplasia, Hay-Wells Syndrome, wounds

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Bacterial wound infections are universally reported within the sparse literature of this disorder. Lesions require intensive wound care.4-6 Acoustic pressure wound therapy (APWT; MIST® Therapy System; Celleration, Eden Prairie, Minn) is a noncontact, low-frequency, nonthermal ultrasound therapy that stimulates fibroblasts critical for wound healing and cleanses and debrides wounds.7 Acoustic energy is delivered to the wound via sterile, saline mist. The effectiveness of APWT has been demonstrated in controlled and case series clinical studies of acute and chronic wounds.8 An in vitro study showed that APWT destroyed bacterial cells.9 Case series evidence suggests that APWT reduces wound-related pain.10 This case series describes the outcomes of APWT for the treatment of extensive and severe scalp wounds with a 3-year history in 3-year-old twins with HWS.

Case Report

Non-Hispanic Caucasian 3-year-old fraternal twins (male and female) were referred to the authors' clinic in February 2008 for worsening wounds. They presented with scalp wounds reaching from ear to ear and from frontal to parietal lobe, with a crusty build-up around the perimeter (see Figure 1a,b). The boy's wounds extended to one side of his face and both ears. The girl had more extensive and recalcitrant wounds. The wound beds were pale pink with thick, viscous, creamy discharge. They were friable and bled easily and profusely. The children cried with dressing changes, suggesting they were experiencing or anticipating pain. At presentation, their dressings consisted of a hydrocortisone 1% cream/bactroban mixture, compounded by their pharmacist, and petroleum gauze. The parents changed the dressings daily.

In addition to scalp lesions, the children had HWS-associated cleft lip and palate, blocked tear ducts, multiple eye and ear infections, light sensitivity, anhidrosis, absence of hair, spontaneous hydronephrosis, dentition abnormalities, and episodes of hypothermia. Both children had surgical repair of the cleft lip, nose, and palate, as well as dental surgery.

At birth, the girl had two small scalp lesions that did not heal and required surgical excision. These wounds remain closed. Shortly after birth, both babies developed a scaly scalp crust that was treated as cradle cap. The crusts thickened and extended, covering the scalps of both babies. Upon debridement, full-thickness wounds were found. These wounds persisted since the children were 3 months (boy) and 5 months (girl) old. Previous management at other wound clinics included multiple topical treatments (ie, foam, oxidized, regenerated-cellulose/collagen with silver, calcium alginate, silver sulfadiazine cream, and biologic tissue matrix), resulting in minimal, short-term improvements.

Informed consent was obtained from the parents to publish their children's photographs and clinical data.
Treatment
The authors selected APWT because other treatments had not provided long-term benefits and because APWT has the combined benefits of decreasing bioburden and promoting healing by stimulating fibroblasts. Initially, APWT was administered once weekly. In consideration of the family’s distance from the clinic, an APWT machine was obtained for their local pediatrician’s office, where treatments could be administered three times weekly, in addition to monthly evaluation at the authors’ clinic. Treatments were 3 to 10 minutes in duration.

Topical treatment was initially silver antimicrobial gel; however, this was not well tolerated, so a broad-spectrum antibiotic/hydrocortisone ointment compounded by the family’s pharmacist was used. The children’s heads were wrapped with roll gauze.

Data were extracted from patient charts from February 2008 to August 2008. Data included number and duration of APWT treatments, wound bed characteristics, and wound dimensions.

The effect of APWT was assessed by evaluating changes in wound bed characteristics and dimensions.

Results
From February 2008 to August 2008, the boy and girl each received 37 APWT treatments. After the boy and girl received five and three treatments, respectively, the scalp crusts were completely debrided, epithelial tissue was observed at the wound edges, and exudate volume decreased. Before the children’s first visit to the authors’ wound clinic, bacterial cultures were positive for *Staphylococcus aureas* and the wounds had purulent exudates. Following initiation of APWT, culture results were negative and clinical signs of infection had resolved.

Substantial improvement for both children occurred during the first 5 weeks of consistent treatment at the clinic (see Figure 2a,b). However, the children missed many treatments due to surgeries, illnesses, and family issues, resulting in deterioration of their wounds (see Figure 3a,b).

In August 2008, the wounds of both children were 70% granulated. The boy’s wounds were substantially improved compared to the girl who had more severe wounds. The boy’s wound decreased in size from 364 cm² to 250 cm² (31.3%); the girl’s wound decreased from 364 cm² to 360 cm² (1.1%).

Treatment with APWT is ongoing. Both children continue to show signs of epithelial progression. During treatments, the children are easily distracted with small toys. Treatments were administered with one parent holding the child on his or her lap and protecting the child’s face from the mist with a washcloth. Considering their young age, the children cooperated well during treatments and did not appear to experience treatment-related pain.
Discussion

As of August 2008, 3-year-old twins with HWS-associated scalp wounds since infancy have received 7 months of APWT treatments. After the first 5 weeks of consistent weekly APWT, the wounds were predominantly granulated with evidence of epithelial progression and no signs of infection. Unfortunately, multiple health and family issues did not allow for either consistent APWT or specialty wound care, resulting in rapid deterioration of the wounds.

The medical literature provides scant information on the treatment of HWS wounds. The authors' experience using APWT as an adjunct to standard wound care in challenging wounds suggested it might be a viable treatment option for HWS scalp wounds. The major benefits of APWT — nonsurgical debridement and reduction of bioburden in the wound bed — were important considerations in the treatment plan for toddlers with extensive, infected, friable wounds.

Conclusion

Results using APWT on HWS-associated scalp wounds illustrate both the potential effectiveness of APWT and the necessity for consistent, long-term treatment of wounds associated with this disorder. Acoustic pressure wound therapy treatments as an adjunct to appropriate wound care may be a valuable option for recalcitrant HWS wounds or other ectodermal dysplasias.

References