

# A NEW THERAPUETIC APPROACH IN THE TREATMENT OF HAILEY-HAILEY DISEASE BY USING POLYMERIC MEMBRANE DRESSINGS\*

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## INTRODUCTION

Familial benign pemphigus also called Hailey-Hailey disease is a rare hereditary blistering skin disease. It typically begins as a painful erosive skin rash at sites of friction that may become thickened, cracked and macerated. There is no cure and treatment is symptomatic.

Polymeric membrane dressings (PMD) have been used with unexpected positive results on two cases. We will describe our findings with the hope that more patients with this condition have the chance to expand their limited choice of treatments.

## Aim

We wanted to look at the;

- Improvement on quality of life
- Reduction of pain
- Effective odor and exudate management,
- Greatest possible independence.
- Improvement of sleep
- Wound healing and maintaining a stable skin situation.
- Reduced risk of recurrence.

## METHOD

Two women with severe long standing skin lesions caused by Hailey-Hailey have been included. They applied the PMD themselves with daily changes, using hydrocolloid strips or silicone tape as fixation. Both patients reported cracked dry wound edges so these were covered by a zinc based barrier creme.

Dietary recommendations as well as a stress free environment was recommended as it is a well known fact that these factors can exacerbate the lesions.

## RESULTS

Patient 1 had large painful lesions under her armpits. She saw a dramatic improvement with reduction of itching and pain within a week, 5 weeks later the lesions had completely resolved. She has continued to use PMD on new lesions and reports that this is the first dressing that has managed to stop the deterioration after a few days. This patient followed her diet restrictions very thoroughly, she was also the one that had the fastest results when it came to healing. Patient 2 had multiple lesions on various body sites. During the first weeks use of PMD there was an increase of lymphatic fluid from her lesions, this normalized after 10 days. Photos taken after 2 months show a huge improvement of all lesions. Both patient reported a dramatically improved sleep situation, waking up only 2-3 times per night instead of several times per hour. Both women reported a reduction of odor and considerably cleaner granulating wounds after a few days.

## DISCUSSION

It is exciting to be able to offer patients with this condition an alternative treatment regime to corticosteroids. Improvement was seen on both patients, ideally we would like to include more patients but as this condition is so rare that will take time to find them.

Both patients make sure they have PMD at home, ready to use as soon as new blistering lesions occur. Pat 1 is especially happy over this solution as her lesions now resolve in a matter of days instead of months.

## Bibliography

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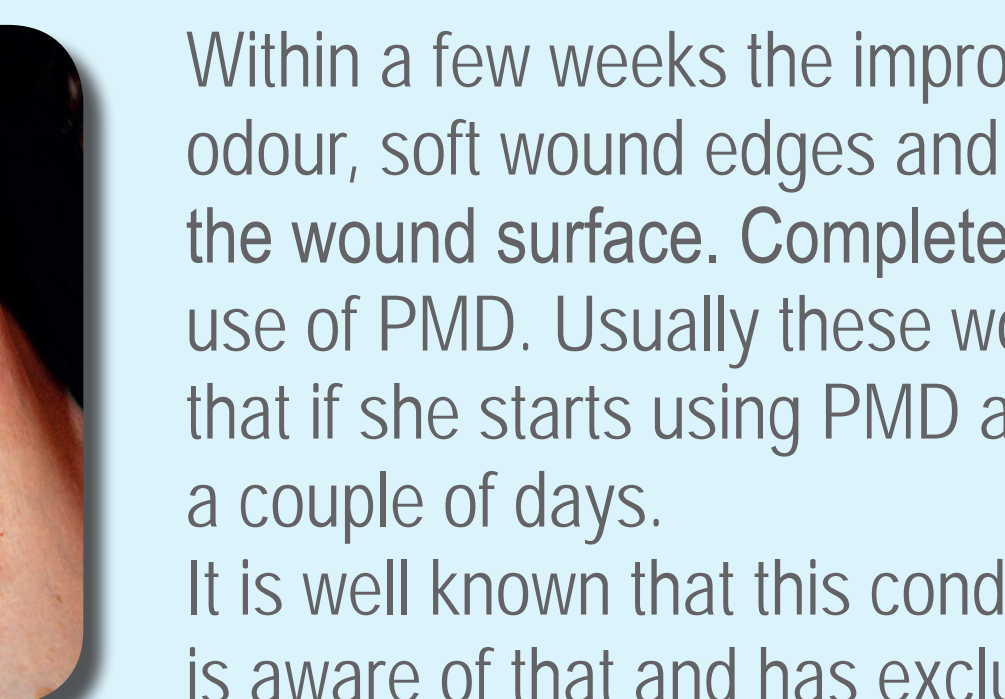
**Pat 1.** Works in the beauty industry. Her father has suffered from Hailey Hailey for 40 years. Mrs M has previously been treated with various cremes/ointments and modern dressings with no effect, the wound continues to deteriorate and enlarge. The wound was a major concern for the patient in her professional life so she was desperate to find a solution that would at least diminish her main problems; bad odour and high exudate level. Having heard about the excellent results with polymeric membrane dressings on Epidermolysis Bullosa, a completely different type of blistering disease, Mrs M wanted to try the dressings on her own wounds which she had been struggling with since her early twenties.



The first couple of days Mrs M only used the dressing at night. Already on the first night she noted that instead of waking up several times an hour she had only woken up twice during the night. The typical, greasy looking scales were dissolving rapidly and the wound surface looked clean and granulating. Based on these results she started to use PMD 24 hours a day.



The armpit is a difficult area to dress as there is much movement in that area. Mrs M used large strips of PMD and fixated them with strips of hydrocolloid which she knew she could tolerate. Since the wound edges had a tendency to dry out and crack she used a protective zinc creme on the edges.



Within a few weeks the improvement was dramatic. Exudate level reduced, no odour, soft wound edges and a with increasing amount of epithelial cells covering the wound surface. Complete wound closure on the 21 July after only five weeks use of PMD. Usually these wounds take 4-7 months to close. She has also noted that if she starts using PMD as soon as new blisters form the rash disappears after a couple of days.

It is well known that this condition is exacerbated by certain diet and stress. Mrs M is aware of that and has excluded all dairy products from her diet.

Familial benign pemphigus also called Hailey-Hailey disease, is a rare genetic blistering skin disease that usually appears in the third or fourth decade. It typically begins as a painful erosive skin rash at sites of friction. As the lesions get bigger the center clears leaving a typical ring shape. Long standing lesions may become thickened, the skin then tends to macerate, become malodorous with painful cracks and wounds.

Heat, sweating and friction often exacerbate the disease, and most patients have worse symptoms during the summer months. There is no cure for Hailey-Hailey disease. Treatment is aimed at reducing symptoms and preventing flares. Common treatments are corticosteroids and antibacterial cremes.

**Pat 2.** Mrs N. is a cheese manufacturer together with her husband. She had her first blistering episode in her early twenties. Almost all skin folds are affected (we chose to show examples from her left armpit). She suffers from a great deal of "burning" pain from her lesions, which also affects the quality of her sleep. As many other patients diagnosed with Hailey Hailey Mrs N. has several food allergies (including dairy products). Dietary restrictions have sometimes been difficult for her to follow. Mrs N. currently receives iron infusions and takes homeopathic medication.



Photo from 12 September shows the status of the wound after 10 days use of PMD. The exudate level was heavy the first 2,5 weeks due to an increase of lymphatic fluid, necessitating dressing changes twice a day. She noted that the pain relief was not as effective when the dressing was saturated. However, the lymphatic fluid decreased after a couple of weeks and with that the malodour.

Like the previous patient, this woman also experienced that her quality of sleep increased, more so when the wound no longer exuded heavily. This had a positive effect on both her professional and personal life.

The PMD was fixated with silicone adhesives due to the fragility of her surrounding skin. Wound edges were coated with zinc creme to prevent the cracked edges that often occur on these types of lesions.



The improvement was not as fast and dramatic as in the first case, maybe due to the fact that she had difficulty in following her dietary recommendations. However, the lesions did improve faster than normal and she now always has a stock of dressings available so she can start the treatment as soon as new lesions appear.