

Surgical control of Hailey-Hailey disease

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Summary—Hailey-Hailey disease is a benign, chronic genodermatosis manifested by recurrent lesions with a predilection for the intertriginous areas. The malodorous, weeping sores are a source of great discomfort to the patient. Medical therapy controls most lesions adequately; however, some are refractory. Experience with excision of skin involved with Hailey-Hailey disease is reported. The best results were obtained in those patients whose wounds were resurfaced with split skin grafts. Primary suture was associated with recurrent disease and residual morbidity. The findings of other isolated reports in the literature are also summarised.

The results show that surgical control of Hailey-Hailey disease is indicated in recalcitrant cases.

Familial benign, chronic pemphigus bears the eponym "Hailey-Hailey disease" after Hailey and Hailey who described the condition in two brothers in 1939. The sufferers are handicapped by malodorous, weeping lesions which occur mainly in the intertriginous areas. Deficient epidermal cell cohesiveness is the underlying cause of this rare genodermatosis. It is controllable but not curable.

The disease affects both sexes, usually appearing in the second or third decade, heralded by the appearance of flaccid vesicles on an erythematous base. The vesicles readily rupture and the denuded areas are soon covered with surface crusts. From the weeping wound, staphylococcus, streptococcus, proteus, pseudomonas or *Candida albicans* are often recovered. Initially the lesions may be intensely pruritic and later become malodorous. The lesions heal after a few months, either spontaneously or with the help of medical treatment. There is residual hyperpigmentation but no scarring. Recurrences occur over many years, usually in the same location (Lever, 1979).

Microscopically Hailey-Hailey disease is uniquely characterised by extensive suprabasal acantholysis. Ultrastructurally the desmosomal complexes are greatly decreased in number. The tonofilaments are detached from their desmosomal attachment plaques and become thickened. Immunofluorescent studies are negative. In clinically normal skin, adjacent to an affected area, the changes are present but less marked (Berger and Lynch, 1971).

This generalised defect in epidermal cell coadhesion means friction may cause acantholytic damage

to the skin. Bacteria also may precipitate the lesions (Shelley and Pillsbury, 1959). Most commonly affected are the groin, perineum, axilla, nape of neck and antecubital fossa. Any area of the body subjected to friction is liable to be affected. We have observed it on the neck due to chafing caused by an arm sling. The disease is worse in the warmer months.

Numerous medical therapies have been advocated to control the active disease. Topical corticosteroids, systemic and topical antibiotics, dressings and baths control most flare-ups. There is, however, a small group of patients who are recalcitrant to these therapies. They are severely socially handicapped by the discharge, dressings and odour. The first report of surgery appeared in 1969 (Biro and Maday, 1969). Since then, there have been several isolated reports of surgical treatment (Table 2; Crotty *et al.*, 1981; Balogh and Freilinger, in press). It is our aim to combine this experience with our own and draw some conclusions about the role of surgery.

Results

There have been nine patients with Hailey-Hailey disease treated surgically in our institution since 1971. One patient, a 54-year-old man, died from a pulmonary embolus three days after excision and skin grafting of the groins. He will no longer be considered in this report (Crotty *et al.*, 1981). All remaining eight patients had long-standing disease. Only one patient was female.

The patients were followed for a minimum of

Table 1 Mayo Clinic Cases

<i>Date</i>	<i>Name</i>	<i>Sex</i>	<i>Age</i>	<i>Length Symptoms (Years)</i>	<i>Area Treated</i>	<i>Technique</i>	<i>Days Inpatient Postop</i>	<i>Length F/U (Years)</i>	<i>Recurrence</i>
1971	R.F.	M	54	25	Groins, perineum, scrotum	Skin graft	14	9	None
1971	J.B.	M	35	20	Groins, scrotum, both axillae	Skin graft	16	8	Minimal in R axillary skin graft
1977	R.W.	M	48	38	Both axillae	Skin graft	22	2	None
1979	W.L.	M	33	9	Groin, scrotum, both axillae	Skin graft groins Suture axillae	18	1	Moderate recurrences in both axillae
1980	A.J.	M	55	37	Groins, perineum	Suture	10	5	Small amount in R axilla, 60% improved
1981	C.R.	F	64	30	Groins, perineum	Skin graft	18	4	None
1986	T.L.	M	53	20	Groins, perineum, R cubital fossa	Skin graft	25	0.5	None. Scrotum improved
1986	M.C.	M	45		Axillae, scrotum	Skin graft, suture			Too recent to assess

Table 2 Other Related Cases

<i>Date</i>	<i>Name</i>	<i>Sex</i>	<i>Age</i>	<i>Length Symptoms (Years)</i>	<i>Area Treated</i>	<i>Techniques</i>	<i>Length Follow-up (Years)</i>	<i>Recurrence</i>
1968	Thorne et al	M	29	2	Groins, scrotum	Skin graft	1	None
1969	Shelley & Randall	F	30	12	Both axillae	Skin graft	4	None
1970	Bitar & Giroux	M	51	34	Perineum, L axilla	Skin graft	2	None
1971	Berger & Lynch	M	51	5	Groin	Skin graft	1.7	None
1974	Rigg	M	26	6	Groin, perineum, part scrotum	Skin graft	0.5	None
1975	Sonck & Rintala	M	35	17	L neck, L axilla, R axilla	Skin graft	2	None
1980	Peled et al	M	56	25	Groin, perineum, sacral region	Skin graft	1.7	None
1981	Prose et al	M	51	25	L axilla	Skin graft	1	None
1982	Kauten et al	M	54	7	Perineum, scrotum	Suture	1.5	None

6 months; all were improved. The six patients who were grafted remained free of morbidity in the region treated.

All patients were admitted for intensive medical therapy prior to surgical treatment. This consisted of regular baths, frequent dressings, topical corticosteroids and systemic and topical antibiotics according to the wound cultures. Often, 2 weeks were required before the wound was clean enough to embark upon excision and grafting.

Reference to Table 1 will show that the groin was

most frequently treated, on six occasions. Three patients had their scrotums resurfaced; one had an area of scrotum excised and closed directly. Axillae were excised bilaterally in four patients. Most wounds were closed with split skin grafts applied either primarily or delayed. Primary suture of the groin/perineum was done in one patient, suture of the axillae in another and suture of the scrotum in a third.

Patients were discharged from hospital when they could manage the wounds at home, even if

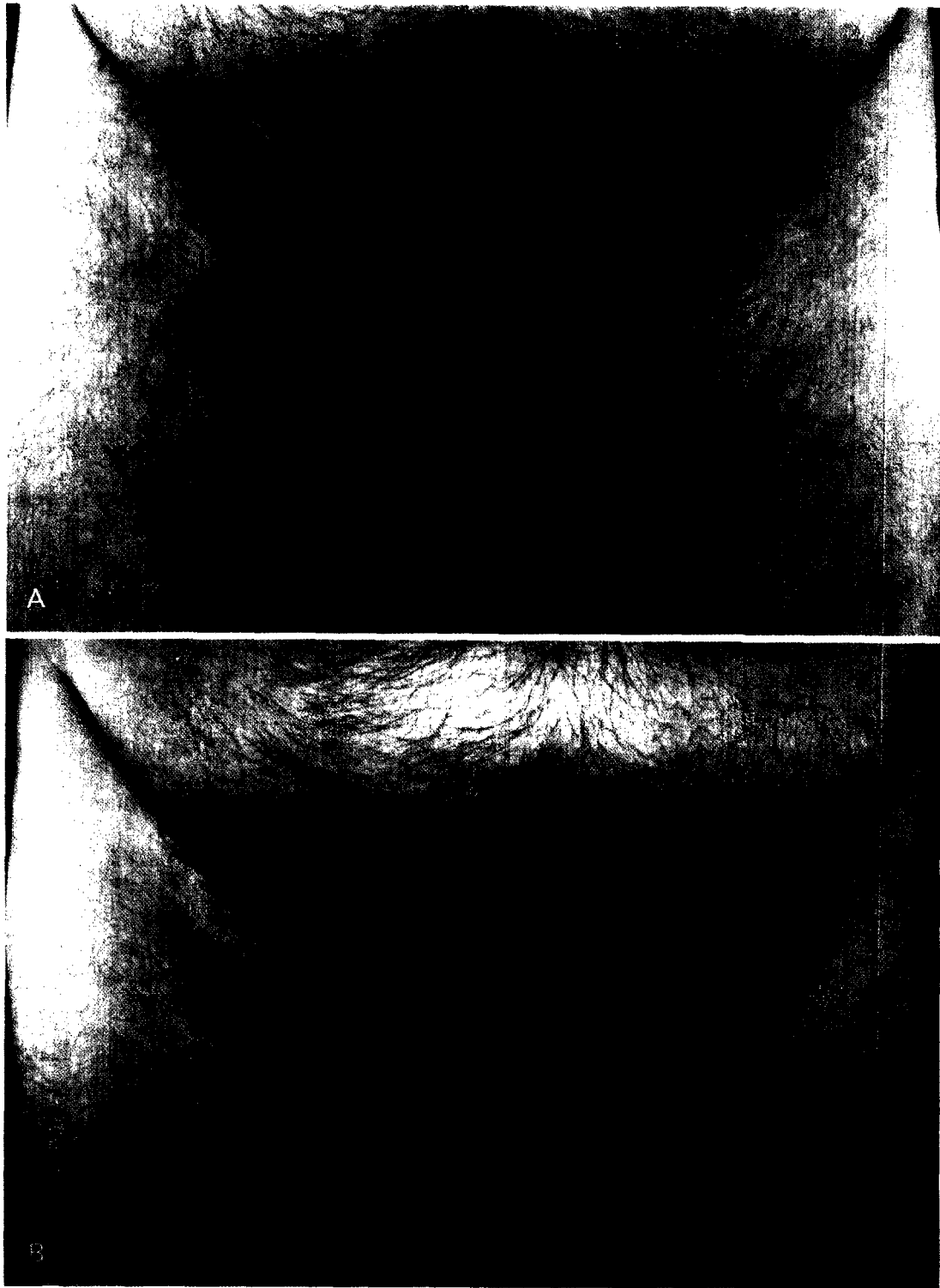


Fig. 1

Figure 1—(A) Extensive involvement of groins, medial thighs and scrotum. (B) Appearance 6 months after excision and skin grafting of groins and medial thighs.

they were not quite healed. Patients have been reviewed a minimum of 6 months after surgery.

Three patients were free of disease in the area treated. It is common to find non-troublesome crusts at the edge of the grafts. Patient T.L., who had widespread disease between the legs, had all skin in the region excised, except the scrotal skin, which was involved with disease. The right antecubital fossa was also resurfaced with a split skin graft. At follow-up, the residual disease on the scrotum was much improved after resurfacing of the surrounding area. This was controllable and acceptable to the patient (Fig. 1). One patient, J.B., had a small recurrence on the axillary graft which could be readily controlled with medical therapy.

Both patients with areas which were sutured directly suffered recurrence. A.J. had slow healing of his groins and had recurrence which was controllable, but still caused morbidity. He felt he was 60% improved. W.L. still required regular therapy for his axillae and felt these areas were little improved. His grafted groins, however, remained free of disease. A recent case is too early to assess.

Table 2 shows results of others reported in the English language literature. The report by Kauten *et al* (1982) describes primary wound suture with no apparent recurrence. The rest of the reports show good results with grafting.

Discussion

All previous studies report excellent results with grafting. This was confirmed in our study.

In this series, the significant recurrences occurred in those areas which were primarily sutured. The in-patient stay may be less and there is no skin graft donor site to worry about. However, the epidermal defect is universal and new lesions will likely manifest if the conditions are suitable. Friction of the skin, particularly in a moist environment, will provide the best potential for lesion development. The maceration may not cause the lesions, but favours growth of bacteria and fungi. It is not logical to bring skin, with the potential to develop florid lesions, into the intertriginous areas. Split skin grafts do not possess sweat glands and therefore provide a drier milieu. The case reported by Kauten *et al* (1982) revealed no symptoms after 16 months, although disease could develop subsequent to this. In fact, there does seem to be some disease adjacent to the groin suture line in their patient in the

6 months photograph. Thus, the grafting approach is recommended.

We have found, like others (Berger and Lynch, 1971; Prose *et al*, 1981), that flare-ups may occur in atypical areas during the surgical procedure. Patient T.L. suffered new lesions postoperatively on his back and under cardiac monitoring electrodes. These areas settled with therapy once the causative factors were removed.

Intensive preoperative lesion therapy is recommended to remove crusts and lower the level of infection. Particularly, B haemolytic streptococcus should be eliminated prior to grafting.

It is reasonable to omit excision of scrotal skin when dealing with widespread inguinoscrotal disease. This lessens the size of the wound to be grafted and the scrotum settles with the drier environment which has been created.

Axillary wounds can be adequately immobilised after grafting and do not present such a great problem, although the method of immobilisation may cause lesions elsewhere. The groins, however, are impossible to immobilise and the grafts are at risk of infection due to the moist environment. For this reason the delayed open grafting technique is advised, with the legs spread apart. This obviously necessitates a private room for the patient.

Finally, it is wise to remember that these patients may previously have had radiation therapy to the affected areas. One report revealed basal cell carcinoma developing in such a circumstance (Bitar and Giroux, 1970).

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Received 18 December 1986.

Accepted 6 April 1987 after revision.