Surgical Wound Healing in patients with Epidermolysis Bullosa

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Background

Epidermolysis Bullosa (EB) is heterogeneous group of rare heritable disorders characterized by blistering of the skin with minor trauma¹. Despite limited evidence to suggest patients with EB have more postoperative wound complications than the general population, we have noted reluctance among some surgeons to operate on these patients due to concerns regarding wound healing. We consequently designed this study to investigate postoperative wound healing in patients with EB.

Figure 1. A healed postoperative scar on the left breast of a female patient with Recessive Dystrophic Epidermolysis Bullosa (RDEB) – note the dystrophic nails associated with disease

Methods

Living patients with confirmed EB were recruited from the Australian National Diagnostic Laboratory Database for EB² and the Australasian EB Registry³. Patients were posted and emailed the ‘Surgical Wound and Scar Healing in EB’ questionnaire which contains twenty-two questions about a patients experience of having surgery (figure 2). The study was approved via a full board review by the Bellberry human research ethics committee, application number 2015-05-369.

Results

351 patients were sent the questionnaire. Forty-six reported a total of 94 surgical procedures. The most frequent surgical procedure was the excision of a skin lesion, followed by musculoskeletal surgery, a caesarean section or an appendectomy. Five patients reported blistering at the surgical wound site after seven surgeries. Three out of these five had EBS generalised severe, one had EBS generalised intermediate and the other had DDEB, none had EBS localised. Four patients reported four postoperative wound infections and one reported postoperative wound dehiscence. Twenty patients had 34 postoperative wounds which they felt healed slower than someone without EB and 30 patients had 55 postoperative wounds which they felt healed at the same rate as someone without EB. After 67 surgeries, the postoperative scar was reported to heal flat and after 18 it was reported to heal with keloid scarring.

Figure 3. A patient with RDEB who has required multiple surgeries for complications related to EB

Discussion

It is unlikely that patients with localized EB simplex will develop blistering at the postoperative wound site, but about a quarter of patients with generalized EB may develop blistering which is likely to interfere with wound healing. Postoperative wound infections do not appear to occur any more frequently in patients with EB than in patients within the general population and wound dehiscence is uncommon in patients with EB. Postoperative wounds may heal at a similar or slower rate in patients with EB compared to those within the general population and patients with EB may have a propensity to develop keloid scars.

Despite the inherent limitations of this study being based on a survey, with this data we feel clinicians should be more confident in referring patients with EB for surgery and surgeons reassured about postoperative wound healing and complications.

References

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Surgical Wound and Scar Healing in EB

Date: _____ Name: _____
Gender: _____ D.O.B: _____

1. What type of EB do you have?
☐ EB Simplex ☐ Dominant Dystrophic EB
☐ Recessive Dystrophic EB ☐ Junctional EB

2. How many surgical operations have you had that have involved cutting through the skin?
☐ 0 - please skip to question 18
☐ 1 - continue to question 3
☐ More than 1 - please fill out a separate form for each operation (even if for the same site, do not include dental work or eye surgery)

3. What was the operation for? What did it involve?

4. Was the operation?
☐ Planned (elective) - e.g. feeding tube inserted into your stomach, planned caesarean section, removal of a skin cancer or correction of a bone deformity
☐ An emergency with no damage or contamination of the skin - e.g. having your appendix out, having an emergency caesarean section
☐ An emergency with damage or contamination to the skin - e.g. drainage of an abscess or an open fracture
☐ Other - please provide details

5. How old were you when you had the operation?
☐ Child (0-12 years) ☐ Teenager (13-17 years)
☐ Adult (18-64 years) ☐ Senior (65 years or over)

6. Where was the skin cut for the operation?
☐ Face ☐ Scalp or neck
☐ Chest ☐ Breasts
☐ Abdomen ☐ Back
☐ Arms ☐ Hands
☐ Groin ☐ Buttocks
☐ Legs ☐ Feet
☐ Other - please provide details

7. How big was the wound?
☐ Very small (<7mm) - e.g. a skin punch biopsy
☐ Small (7mm-2cm) - e.g. key-hole surgery
☐ Medium (2-6cm)
☐ Large (>6cm) - e.g. abdominal or chest surgery

8. How was the wound closed?
☐ Sutures or staples on the outside
☐ Dissolvable internal sutures
☐ The wound required a skin graft
☐ Other - please provide details

9. Did you develop blisters at the wound site after the surgery? (not related to bandages, tapes, cannulas etc.)
☐ No
☐ Yes - if so do you think they interfered with the wound healing? ☐ Yes ☐ No

10. Did you develop blisters/wounds due to bandages, tape or IV cannulas etc. while in hospital?
☐ No
☐ Yes - please provide details

11. Did you develop any postoperative complications?
☐ No
☐ Yes - please specify:
☐ Wound infection requiring antibiotics
☐ Wound dehiscence (stitches/sutures/staples broke and the wound opened up)
☐ Other - please provide details

12. How long did the wound take to heal?
☐ Less than one week
☐ One to two weeks
☐ Two weeks to one month
☐ One month to three months
☐ Three months to a year
☐ It never healed (I still get problems with the skin breaking down where the operation was)

13. Comparing the healing of the wound to someone who didn't have EB, do you think the wound healed?
☐ Slower
☐ Faster
☐ The same

14. How did the scar heal?
☐ Flat ☐ Keloid (raised)
☐ Hypertrophic (thickened) ☐ Atrophic (sunken)

15. What color is the scar now?
☐ Lighter than my skin ☐ Darker than my skin
☐ Red ☐ Same colour as my skin
☐ Other - provide details

16. What is your opinion of the scar now?
(very ugly) 0 1 2 3 4 5 6 7 8 9 10 (perfect)

17. Do you have any advice for doctors looking after patients with EB in the hospital after having surgery?

18. Have you ever had a tattoo(s)?
☐ No - please skip to question 22.
☐ Yes - what parts of the body?

19. Did you get blisters/wounds at the site of the tattoo before getting the tattoo(s)?
☐ Yes
☐ No

20. When you had the tattoo(s) did it cause blistering?
☐ Yes
☐ No

21. Do you get blisters/wounds over the tattoo now?
☐ Yes
☐ No

22. Thank you for completing the questionnaire. Do you have anything else you want to add that may be relevant?

Figure 2. The ‘Surgical Wound and Scar Healing in EB’ questionnaire