

# Hand Surgery for EB

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# Overview

- Presentation
- Timing
- Surgical Options
- Post Surgical Care and Outcomes

Presentation













# Timing


- Support
- Patient goals
- Access to therapy
- Age
- Joint status

REVIEW

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# Hand surgery and hand therapy clinical practice guideline for epidermolysis bullosa

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## Abstract

**What is already known about this topic?:** Epidermolysis bullosa (EB) causes blistering and scarring of the hands resulting in contractures fused web spaces and altered function. Surgery is needed to release contractures and web spaces and hand therapy is essential to maintain results, approaches for both differ.

**What does this study add?:** These guidelines aim to provide information on the surgical and conservative therapeutic hand management of children and adults diagnosed with EB. They are based on available evidence and expert consensus to assist hand surgeons and therapists in decision making, planning and treatment. They highlight the importance of a holistic multidisciplinary team (MDT) approach, where patient priorities are paramount.



# Findings and Recommendations

- Most families are satisfied with outcomes after hand surgery
- Hand surgery improved patient function with moderate to severe EB
  - Improvement is temporary with recurrence in 1-2 years
  - Better outcomes at younger ages
  - Joint deformities mean worse outcomes
- Surgical decisions should include patient, carer, surgeon and therapist
- Anesthesia
  - Use an experienced team
  - Regional blocks

# Surgical Options

- Removing the mitten
- 1<sup>st</sup> webspace only versus whole hand
- Joint fixation
- Skin grafting
- Dermal substitute
- Gene therapy











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# Release of pseudosyndactyly in recessive dystrophic epidermolysis bullosa using a dermal regeneration template glove: the Foggia experience

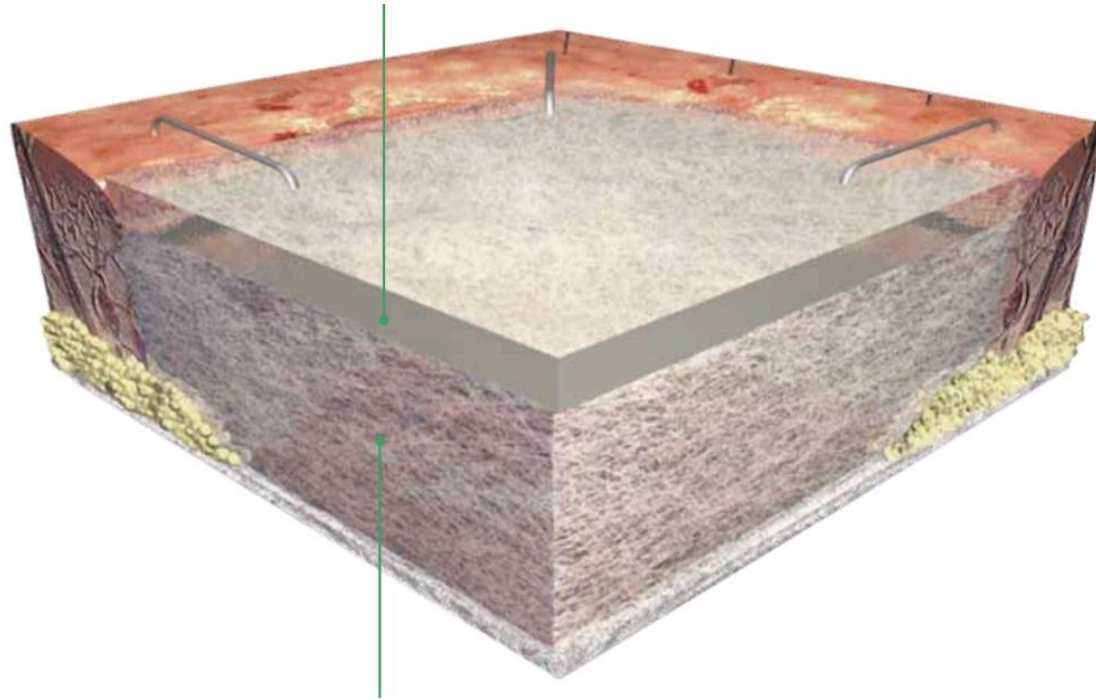
Fedele Lembo<sup>1\*</sup>, Domenico Parisi<sup>1</sup>, Liberato Roberto Cecchino<sup>1</sup>, Francesco Ciano<sup>1</sup>, Alessandro Innocenti<sup>2</sup> and Aurelio Portincasa<sup>1</sup>





### Silicone layer

- Enables immediate wound closure
- Controls fluid loss
- Provides mechanical protection
- Provides a bacterial barrier
- Water vapor transmission rate similar to that of normal skin



### 3-Dimensional matrix layer

- Cross-linked collagen and glycosaminoglycan
- Functions as an extracellular matrix
- Promotes cellular growth and collagen synthesis
- Biodegrades while being replaced by autologous dermal tissue

**Table 2 Outcomes of the two types of surgical approaches for hand pseudosyndactyly**

	Group A	Group B	<i>p</i> -value
Healing times (days, mean $\pm$ SD)	46.4 $\pm$ 3.4	24.6 $\pm$ 2.9	0.008
Hospital stay time (days, mean $\pm$ SD)	29.6 $\pm$ 1.6	3.6 $\pm$ 0.8	0.021
Discomfort for the patient (through VAS-scale, mean)	7.6	0.6	0.034
Free-recurrence interval (months, mean $\pm$ SD)	19.1 $\pm$ 5.3	31.1 $\pm$ 4.7	0.045
Follow-up range (years, range)	3.7–8	1.5–9	–
Major complications	0	0	–

Group A: Patients underwent surgical release and dressing with vaseline gauze

Group B: Patients underwent surgical release and dressing with Integra®











# Post Surgical Care and Outcomes

- Dressing changes
- Splinting
- Hand therapy



















- Dressing holds the hand open
- Changed at home 2-3 days
- Thermoplastic splint by therapist





















# Conclusions

- Better results without joint deformities
- Improved function is possible even with severe disease
- Recurrence is expected
- Patient and caretaker commitment is critical
- Hand therapy, dressings, and exercises help prolong outcomes



Thank You

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