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Surgical management of hand deformity in *Epidermolysis Bullosa*: our experience in a case series of pediatric patients

Gestione chirurgica della deformità della mano in Epidermolisi Bullosa: la nostra esperienza in una serie di casi di pazienti pediatrici

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Summary

In this study we present 2 patients for a total of 2 surgically treated hands. Data were collected between 2017 and 2023 at Azienda Universitaria Policlinico di Modena (Modena, Italy). The postoperative follow-up period ranged between 12 months and 18 months, with an average duration of 15 months. The procedure performed on these patients involved a first web release for the thumb and/or pseudosyndactyly release for the remaining digits and/or the surgical treatment of camptodactyly with Malek cutaneous approach. Acellular dermal substitute (Matriderm®) was used to cover the remaining commissures, digits, and the remainder of the hand. Postoperative rehabilitation ensued. The patient's medical history, physical examination findings, and treatment are described. The outcome of surgical release and rehabilitation are discussed. Results: Long-term results are encouraging, demonstrating maintenance of functions.

Key words: epidermiolysis bullosa, hand surgery, hand deformity, surgical management, anesthetic management, surgical technique

Riassunto

In questo studio presentiamo 2 pazienti affetti da Epidermiolisi Bollosa autosomica recessiva distrofica, per un totale di 2 mani trattate chirurgicamente. I dati qui presentati sono stati raccolti tra il 2017 e il 2023 presso l'Azienda Universitaria Policlinico di Modena (Modena, Italia). Il periodo di follow-up postoperatorio variava tra i 12 mesi e i 18 mesi, con una durata media di 15 mesi. La procedura eseguita su questi pazienti ha previsto l'apertura della prima commissura per il recupero di motilità del pollice e/o la correzione della pseudosindattilia per le restanti dita e/o il trattamento chirurgico della camptodattilia con l'approccio cutaneo di Malek. È stato utilizzato un sostituto dermico acellulare (Matriderm®) per coprire perdite di sostanza residuate dall'apertura delle commissure. I

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Conflitto di interessi
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pazienti qui destritti sono stati indirizzati verso un ben definito percorso di riabilitazione postoperatoria. Vengono descritti la storia medica del paziente, il trattamento e i risultati osservati. Si discute il risultato del release chirurgico e della riabilitazione. Risultati: i risultati a lungo termine sono incoraggianti, dimostrando il mantenimento delle funzioni recuperate (es funzione di pinza).

Parole chiave: *epidermolisi bollosa, chirurgia della mano, deformità della mano, gestione chirurgica, gestione anestesiologica, tecnica chirurgica*

Introduction

Epidermolysis Bullosa (EB) was first described as 'erblichen pemphingus' by Von Hebra in 1870 [1]. It is a group of rare skin conditions, that result in skin and mucous membranes fragility, which can be an acquired or inherited disorder.
Based on the site of formation of the blisters, it is possible to identify four types of EB:

- EB simplex (EBS);
- Junctional EB (JEB);
- Dystrophic EB (DEB);
- Kindler syndrome (KS).

Each type can be further classified based wether on the mode of transmission, or on the clinical phenotype or on the immunofluorescence, structural and molecular pattern [2-3].
Alternatively they can be classified based on the splitting site:

- epidermolysis bullosa simplex: epidermis;
- junctional epidermolysis bullosa: within the lamina lucida of the basement membrane zone;
- dystrophic epidermolysis bullosa: within the dense sublamina;
- Kindler syndrome: variable [4].

Hallmark of the EB is the blistering of the skin and mucous membranes. Blisters occur due to traumas, which lead to the development of chronic ulcers, often sparsely cicatrising, with propensity to infections, pain and itch.
EB severity can range from mild to fatal. In mild cases the patients present superficial blisters, mostly located on the hands and feet, which do not influence their life span. On the other hand, in severe cases patients risk death in the first postnatal period due to infections or electrolyte imbalance. Furthermore, few types of EB have the propensity to develop aggressive squamous cell carcinomas which can quickly metastasise leading to premature death [5].

Hand deformity in patients affected by EB

As explained above all EB types hit the hands, however the patients that need to undergo surgery and a therapeutic plan are the ones affected mainly by Dystrophic epidermolysis bullosa (DEB).
The hand is a delicate region, strongly prone to the development of blisters, ulcers and scars due to simple tangential forces and daily friction [6-8].
Common deformities include thumb adduction contractures, digit pseudosyndactyly, flexion contractures of finger inter-phalangeal joints (IPJs) and metacarpal phalangeal joints (MCPJs) and wrist, occasional extension contractures of MCP joints from dorsal scarring.
A mitten deformity develops when the hand becomes encased in an epidermal cocoon. In Recessive Dystrophic Epidermolysis bullosa (RDEB) the risks of this developing are 98%by the age of twenty [6-9].
All hand structures may be affected by the disorder. Cutaneous involvement results in dermal fibrosis, pseudosyndactyly, contractures, atrophy of finger and thumb tips, nail loss and dermal cocooning.
Musculotendinous involvement results in flexor tendon shortening and intrinsic muscle contractures. IPJs and MCPJs flexion causes collateral ligaments to contract and become fibrotic overtime. Constant abnormal stress and deforming pull on joints by contracting scar tissue causes destructive joint changes and subluxation. All web spaces become obliterated progressing to digit tips. Advanced hand deformity results in functional impairment including loss of fine motor manipulation [8].

Surgical indications

The evidence as to whether surgery improves hand function is not clear, due to the lack of large, controlled studies. However, it is proven by most data that surgery gives a chance of improval in hand function in DEB cases both in severe and moderate stages of deformity [8].
Obtaining a functional thumb-index pinch is the main goal of surgical treatment. The release of the thumb adduction contracture produces the most dramatic improvement in patients, together with surgical treatment of camptodactyly. The main goal would be the release of all fingers, if possible.
Surgery and release of pseudo syndactyly allow independent finger motion and improve aesthetic appearance of the hands. In particular, surgery in children may help prevent developmental and motor delay and hand atrophy [11,12].
However, it needs to be taken into account that the positive effect of surgery is not constant, showing a Gaussian distribution curve, rising with healing, and decreasing in time with recurrence.

The decision to operate, therefore, needs to be taken based on several factors:

- good health status of the patient;
- rapid progression of hand contractures and pseudosyndactylies;
- loss of manual function;
- the patient's or parents' request;
- the effect on the psychosocial development of the patient.

It is preferred to operate initially before the patient turns eleven years old. It is in fact proven that the correction of contractures in children often leads to more satisfactory results rather than in adolescents or adults, due to the fact that joint deformities in the lasts present themselves as too difficult to be corrected completely [14-16].

Anesthesia

Considering the gravity and rarity of the disorder, it is strongly recommended that an experienced EB nurse is present during surgery, to highlight specific problems: previous anesthetic, airway, vascular access, or medical problems, and provide comfort and support to the individual.

In particular, their role is to advise and educate the anesthetic and theatre team on skin care, and what procedures to avoid (adhesive dressings, safe handling) [8].

Taking into account anestahesia, the type used appears mainly related to age. Younger individuals are more likely to undergo General Anaesthetic (GA), and older individuals Regional Anaesthetic (RA) ± intravenous ketamine or sedation. Given that mouth opening tends to worsen with age, RA is increasingly preferred to avoid risks to the airway [8].

In children when used flunitrazepam and ketamine and either an axillary or supraclavicular block, brachial plexus blocks, and ketamine were reported [17].

Pre-operative care

The patient is carefully placed on an operation table that must be well prepared to prevent pressure ulcers. Large padding should be used at the pressure zones, and repositioning of the patient during the procedure should be avoided. The skin is disinfected using a "buffering" technique or a spray of the preparation, preferably in nonalcoholic form [8].

Anesthetic creams such as EMLA (lidocaine-prilocaine) are usually used to provide local skin anesthesia before intravenous cannulation in children. Ultrasonography can be useful for vein canulation or blood drawing [8].

No adhesive tapes are used, soft bands are usually preferred. Skin fragility also poses an infection risk. Wound care management is done according to the DEBRA guidelines. Finally, the management of pain pre- and postoperatively can be challenging in these patients due to the presence of chronic pain, reliance on analgesics, and physical stress intolerance in some [8].

The anesthesiologist chooses the type of anesthesia most suitable for the patient. General anesthesia must be avoided. There exists a risk of complex intubation due to the presence of microstomy, fragility of the oral and nasal mucosa and laryngotracheal stenosis in some. Laryngeal masks are to be avoided. In our study, intravenous sedation (ketamine) coupled with locoregional anesthesia is the method of choice. Intravenous sedation and ketamine usually have the advantage to not cause oropharyngeal trauma. Ketamine, however, can increase oropharyngeal secretions that require special attention and monitoring [8].

Perioperatively, the antibiotic most often given to patients is Augmentin® (amoxicillin + clavulanic acid). When hardware is placed (i.e., Kirschner wires), antibiotics of the cephalosporin family (first generation) are used. Antibiotics are not indicated postoperatively unless there are signs of infection [8].

Post-operative care

Fundamental in the first few weeks following surgery is to protect the surgical site by caring for the skin. If the skin was grafted, this must be closely monitored for integrity. Timing of skin reepithelization varies but has been reported to be achieved around 14-35 days postop [11-18].

It is strongly recommended that the first dressing layer, in contact with the skin, consists of a non-adherent gauze such as Vaseline Petrolatum or Hollister Restore, or a soft silicone or foam product, such as Mepilex Lite, Mepitel or Mepilex Transfer. The first dressing layer should be arranged or cut in a fashion that covers all surgically affected skin maintaining web spaces.

Under specific indication of the surgeon, the silicone or foam layer may be covered with a thin layer of Vaseline in the manner of "buttering bread."

The base layer is held on by one-inch gauze wrapping in a secure, overlapping "boxer's wrap" fashion, including web spaces and digits. It is possible, if preferred by the patients, to wear a soft stockinette over dressings, with the wrapped thumb and fingers free to move [10,13,17,19-22].

In addition, it is advised that post-operative hand orthoses/splints are worn to maintain the surgical gains and help delay contracture recurrence, as displayed by most articles. This should start as soon as individuals can tolerate the fabrication process, which may be while the hand is still fully dressed.

However, timing also depends on whether fixation (i.e., Kirschner wires) is used. The wounds may leak onto the splint/orthosis, so lining and strapping materials should

allow easy replacement or cleaning with soap and water [10,13,17,19-22].

It is advisable to fabricate a hand or forearm based resting splint/orthosis including the fingers and thumb. The splint/orthosis should hold the fingers and thumb in maximum passive extension and abduction, as tolerated by the individual. It should be lined or padded to cushion the hand and protect the skin. Strapping should also be as soft as possible [10,13,17,19-22].

Where only the first web space is released, the thumb should be positioned in abduction with a silicone elastomer putty spacer, held in place by a thermoplastic splint/orthosis or careful wrapping.

Based on past experiences, we recommend full time splint/orthosis wear from week three or four post-op, removing only for light activity and exercise [23]. Only from two to four months postop individuals may transition to use only at night [13,22].

Individuals should begin to use their hands for function approximately four to five weeks post op.

Material and methods

This was a retrospective review with data collected from the archives of Azienda Ospedaliera-Universitaria Policlinico di Modena. The data reviewed were collected between 2017 and 2023.

Patient profiles

This study includes 2 patients with RDEB who underwent surgical releases of either thumb adduction [1] or the surgical treatment of camptodactyly of II and III IFP [1] with Malek cutaneous approach, including the use of dermal substitute Matriderm.

One female and one male patient, with an average age of 14 years (range: 13-15 years) were treated by a single operator (sg). Surgery was performed on 2 right hands. A total of 2 operations took place, with 2 hands having undergone a single procedure (100 %). One patient underwent previous surgeries at other institutions.

A total of 2 hands had Matriderm 1 mm placed (100%). The postoperative follow-up period ranged between 12 months and 18 months, with an average duration of 15 months.

Anesthesiology Management in EB patients

All EB patients have been evaluated by a highly skilled anesthesiology team given the complexity of this patient population and scheduled well in advance to facilitate multidisciplinary coordination and completion of preoperative testing. The extent of cutaneous involvement has been documented, with particular attention to the airways, eyes, and upper extremities. Patients with severe EB are expected to have limited mouth opening with smaller interincisive distance, dental caries, and limited neck extension from contractures, which is im-

Table I. Clinical characteristics and surgical treatment performed.

Patient number	Sex	Diagnosis	Previous treatment	Age when operated (y)	Hand operated	Artificial dermis	Kirschner wire	Anesthesia	Follow-up (months)	Comorbidities
1	F	Pseudosyndactyly: simple and complete	Yes	15	R	Matriderm	Yes	Locoregional	18	Esophageal strictures
2	M	Camptodactyly IFP II-III fingers	No	13	R	Matriderm	Yes	Locoregional	12	/

Table II. Anesthetic characteristics and corresponding management.

Paz (age)	Difficult airways	Regional Block technique	Perioperative Sedo-analgesia protocols	Postoperative analgestia	Complications
Pz 1 15 anni 37 kg	Yes Interincisive distance < 2 cm	ENS - Ultrasound guided brachial plexus via axillary block tot 95 .mg ropivacaine 0,475%	Midazolam 3mg i.v. Fentanyl 100 mcg iv Propofol 80mg	Paracetamol 500mg/8h Ketorolac 15mg rescue dose	no
Pz 2 13 anni 34 kg	Yes Interincisive distance < 2 cm	ENS - Ultrasound guided brachial plexus via axillary block tot 75 mg ropivacaine 0,475%	Midazolam 6 mg i.v. Fentanyl 30mcg iv	Paracetamol 250mg x3 Nurofen syrup 10ml ogni 88hy	no

portant for difficult airway planning. The presence of hoarseness or inspiratory stridor may suggest evaluation for laryngeal involvement. For hand surgery procedures, a combination of regional anesthesia and sedation was chosen to avoid potential complications associated with airway management.
Careful planning and precautionary measures must be taken to minimize the risk of complications.
Standard anesthesia monitors have been safely used in patients with EB with nonadhesive pulsoxymeters and a special gel pad that provides a conductive, nonadhesive barrier between the electrode and the skin.
Blood pressure cuffs have been placed over cotton undercast padding or existing dressings on the opposite arm.

Surgical technique
In this study, hand surgery is performed under locoregional (with or without sedation) anesthesia.

Hand release
Case 1. A 15-year-old DEB-diagnosed girl, who had previously undergone a first web release for the right thumb and pseudosyndactyly release presented recurrence of thumb adduction contractures, digit pseudosyndactyly and flexion contractures of finger inter-phalangeal joints (IPJs).
Under LR anesthesia, a cutaneous incision was performed on the dorsal aspect of the first interdigital space, between the 1st and 2nd metacarpal bones, then extended volarly to the base of the thenar region reaching the proximal palmar crease. We proceeded by plans isolating and protecting the nervous vascular structures, with the release of either fibrous tissues or soft tissues of the thenar region, incising the muscle fascia, improving as a consequence the opening of the 1st commissura.
With the aid of the fluoroscope, a 1 mm Kirschner wire is inserted from the distal phalanx of the thumb to the 1st metacarpal locking it also to the trapezium bone. A second 1 mm Kirschner wire is inserted between the 1st and 2nd metacarpal bones. Accurate hemostasis was then performed, washing with physiological solution.

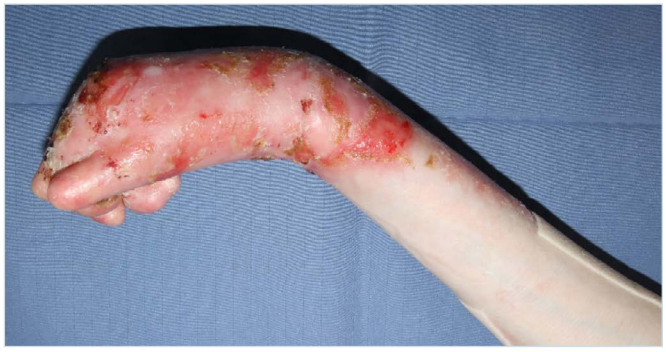

Figure 1. Case 1: pre-operative.

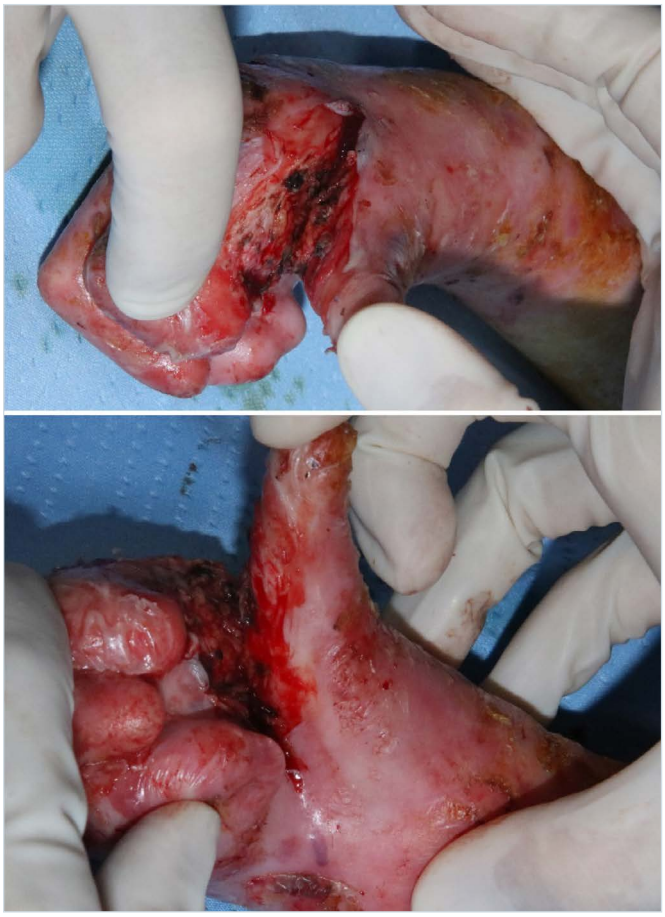

Figure 2. Case 1: post-operative.

In the following month the medication was renewed approximately every 4/5 days, at home or at our centre.
The first Kirschner wire was removed after approximately 10 days due to intolerance.
One month after the operations we proceeded to curettage and cleansing of the right hand. The patient was under sedation. We proceeded to the placement of a non-adherent dressing at the level of the first commissure and removal of ONE K-wire which showed signs of inflammation.
A silicone elastomer-based spacer was then placed over the Mepilex plates to keep the 1st interdigital space open while respecting the synthesis means. A plaster cast is applied to keep the wrist in a neutral position.
About a month later, under local anesthesia and sedation, surgical wound dressing of the right hand was performed and Kirshner's wire was removed.
Case 2. A 13-year-old DEB diagnosed boy was referred to us for camptodactyly of IFP II-III fingers.
Preoperative antibiotic prophylaxis was administered. The patient was placed in supine position with a right upper limb

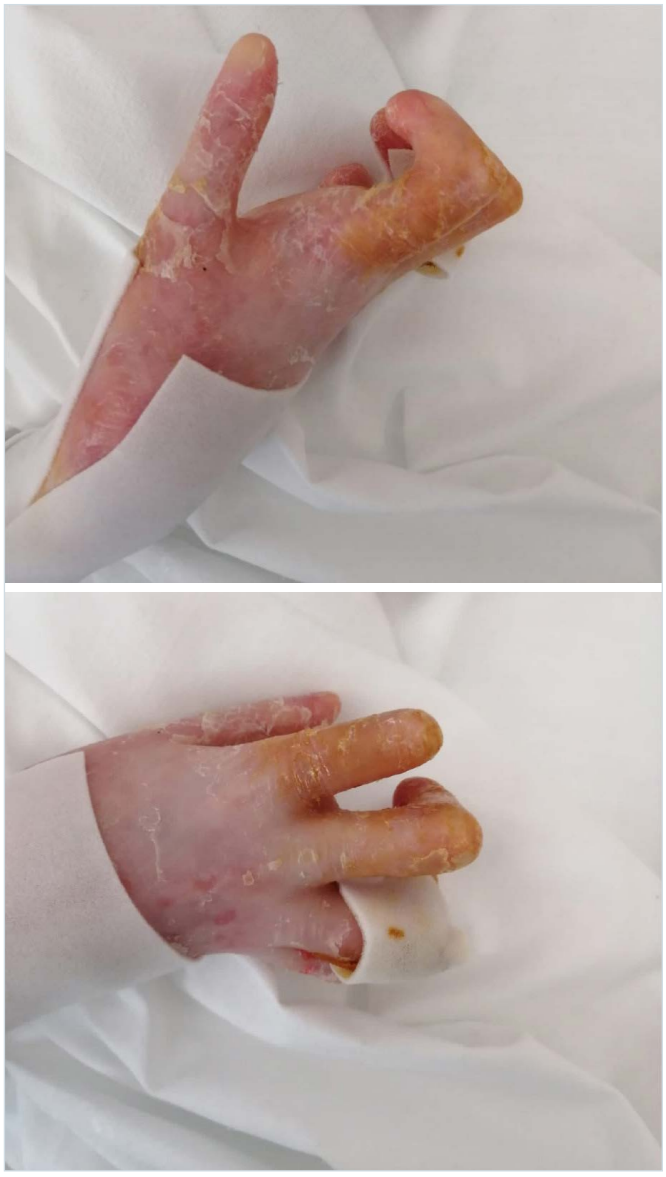

Figure 3. Case 2: pre-operative.

sterile field. Skin elongation plasty with volar flap according to Malek's technique was performed on the 2nd and 3rd finger: we proceeded in stages isolating and protecting the nervous vascular structures, releasing of the subcutis and tendon structures, with recovery of the almost complete passive extension of the fingers. In the operative room, each anatomical structure is assessed before the release to obtain the greatest possible extension. Satisfactory lengthening may require several surgical steps:
Step 1: the volar cutaneous retraction is treated by drawing a proximally based skin flap at the PIPJ, according to the Malek technique.

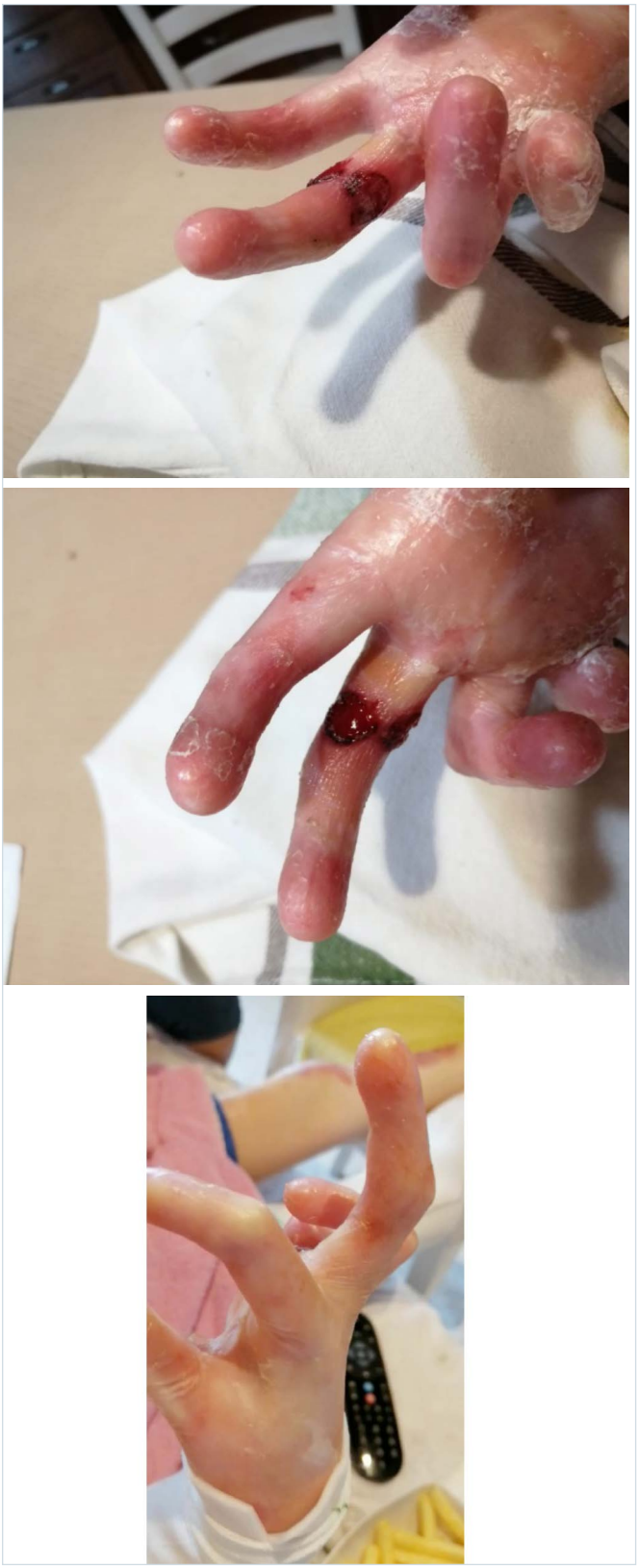

Figure 4. Case 2: post-operative.

Step 2: subcutaneous fibrous structures are released preserving the neurovascular bundles and are used as a coverage for the underlying flexor apparatus. Intra-operative assessment of PIPJ passive extension with extended metacarpophalangeal joint (MCPJ) is performed.
In the following month, the medication was renewed approximately every 4/5 days, at home or at our center.
About a month later, after antibiotic prophylaxis with Cefazolin 1 g, under sedation, surgical dressing of the right hand wound was performed and Kishner's threads removed. We proceed to packing the elastomer. Medication application.

Defects coverage
The technique of skin coverage varies. At the level of the first and second commissure of the hand a dermal substitute was used and our first choice was Matriderm (2 mm). The dermal substitutes is fixed with Rapid Vicryl 4/0 (Ethicon, Johnson & Johnson).
We then placed a below palm plaster valve along with a dressing of Urgotul + Mepilex Lite.

Results
Surgery was performed one hand at a time, to preserve the patient's autonomy as much as possible. Results of our retrospective study demonstrated more postoperative functional improvement in patients with RDEB of lower severity, who had good joint function and rigorous cooperation with postoperative rehabilitation.
The dermal substitute Matriderm promoted tissue re-epithelialization in an average of about 30 days. The Kirschner wires were removed approximately 21 days after the first surgery. At the same time, a maintenance elastomer was set up in the operating room and patients were taught to keep it in place upon discharge.
At home, patients were instructed about the importance of keeping the elastomer in place as much as possible to maintain the opening of the first commissure, as well as the need to exercise the pinching function to aim for maintaining the functionality achieved post-surgery.

Our results are rather in favor of the release of digital pseudosyndactylies and camptodactylies, since based on our case observation, the opening and releasing of both appears to be maintained for over 1 year after the procedure, maintaining digital function.
The release of pseudosyndactylies is in our opinion, necessary when it is possible.
A rapid recurrence can sometimes be the result of noncompliance of the patient with postoperative reeducation. We believe that an appropriate surgical technique followed by rigorous rehabilitation, combined with an interdisciplinary overall management of these patients with RDEB, allowed us to succeed in the optimization of their manual function.
As soon as possible it is fundamental to start physiotherapy sessions, two or three times per week, proceeding as long as improvement is shown. In adults, physiotherapy appears to be more intense and therefore is continued based on the patients' tolerance. The goal remains to be able to overcome postoperative rigidity.
Furthermore during the postoperative physiotherapy evaluations of the second patient (Case 2) the following values concerning the evaluation of strength (using pinchmeter readings in Tip to Tip) and ROM were observed:
Furthermore camptodactyly pre-operative mean extension deficit was 65° (range 35°-100°) and post-operative mean extension deficit was reduced to 16.5° (range 0°-70°).

Conclusion
As sufficiently presented surgery is associated with functional improvement.
Nevertheless, the focus needs to be shifted also on the potential surgical complications (bleeding, infection, very rarely loss of fingers or phalanges), together with pain, loss of hand function while healing, and the risks related to the use of anesthesia.
In addition, even though surgery in children may help prevent developmental and motor delay and hand atrophy, the positive effect of surgery is not constant [11-12].
Some ways to clinically monitor and assess hand function im-

Table III. Postoperative physiotherapy evaluations.

Left					Units (Kg)	Right				
Try 1 2.1	Try 2 2.1	Try 3 2.1	Avge 2.1	CV%: 0	Pinchmeter readings in Tip to Tip	Try 1 1.4	Try 2 1.4	Try 3 1.4	Avge 1.4	CV%: 0
					ROM II finger IPI	FL: 40° EXT: 30°				
					ROM III finger IPI	FL: 40° EXT: 30°				

provement may include dexterity tests such as the nine-hole peg test, and quantitative strength tests such as grip strength or pinch tests. Further studies are certainly needed to evaluate their accuracy and validity as follow-up parameters [24,25].
In fact, improvement in hand function is temporary, with recurrence expected within 1-2 years, 50-53%occurring after 1 year, with approximately 50% requiring further procedures. In one series recurrence was 53 %, another 50 %, with repeated procedures every 2 years to maintain optimal function [7-13-14-15].

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