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Surgical treatment of contracture and syndactyly of children with epidermolysis bullosa

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SUMMARY. **A new approach to the operative treatment of syndactyly and contracture of the hands of children with recessive dystrophic epidermolysis bullosa is described. It is based upon the principle of surgical release of fingers allowing spontaneous epithelialisation of skin wounds without using skin grafts. Nineteen children had operations using this method, with an incidence of recurrence of 53%. This method has the advantage of a short operating time, simple technique and limited trauma.**

Recessive Dystrophic Epidermolysis Bullosa (RDEB) Hallopo-Siemens is the most severe form of epidermolysis bullosa. Blisters appear on the skin and mucous membranes following minor mechanical trauma, and erosions heal with atrophic scar formation. Repeated blistering on hands and feet leads to contractures and fusion of the web spaces. The hand, encased in an "epidermal cocoon", cannot grow and develop properly. The resulting hand deformities and inability to perform fine manipulation adversely affects the child's psychosocial development, providing strong reasons for surgical treatment.

Previous operative approaches have included epidermal "degloving", split thickness or full thickness skin grafting, fixation of fingers with wire devices, and the use of intraosseous K-wires.[1–3] However, there is a high incidence of recurrence of the deformity independent of the surgical method used. The present work describes a modified approach to the surgical correction of hand contracture and syndactyly in children with RDEB.

Patients

In the Moscow Scientific Research Institute of Paediatrics and Paediatric Surgery we have performed 49 operations on 19 children with RDEB aged from 2–14 years. There were 12 girls and 7 boys. All had contracture and syndactyly (Figs 1, 2). Twelve children had very advanced deformities of the "epidermal cocoon" type.

Fig. 1

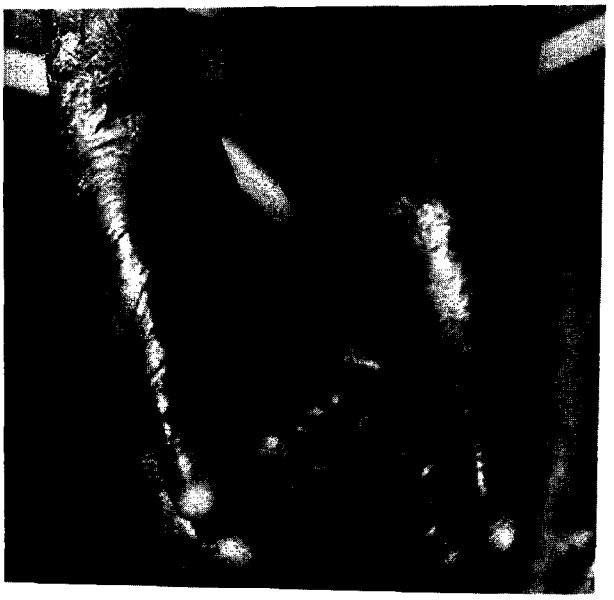

Fig. 2

Figure 1—Dorsal view of hand of 8-year-old girl with contracted fingers and syndactylies. **Figure 2**—Hand of 6-year-old boy with typical deformities of "epidermal cocoon".

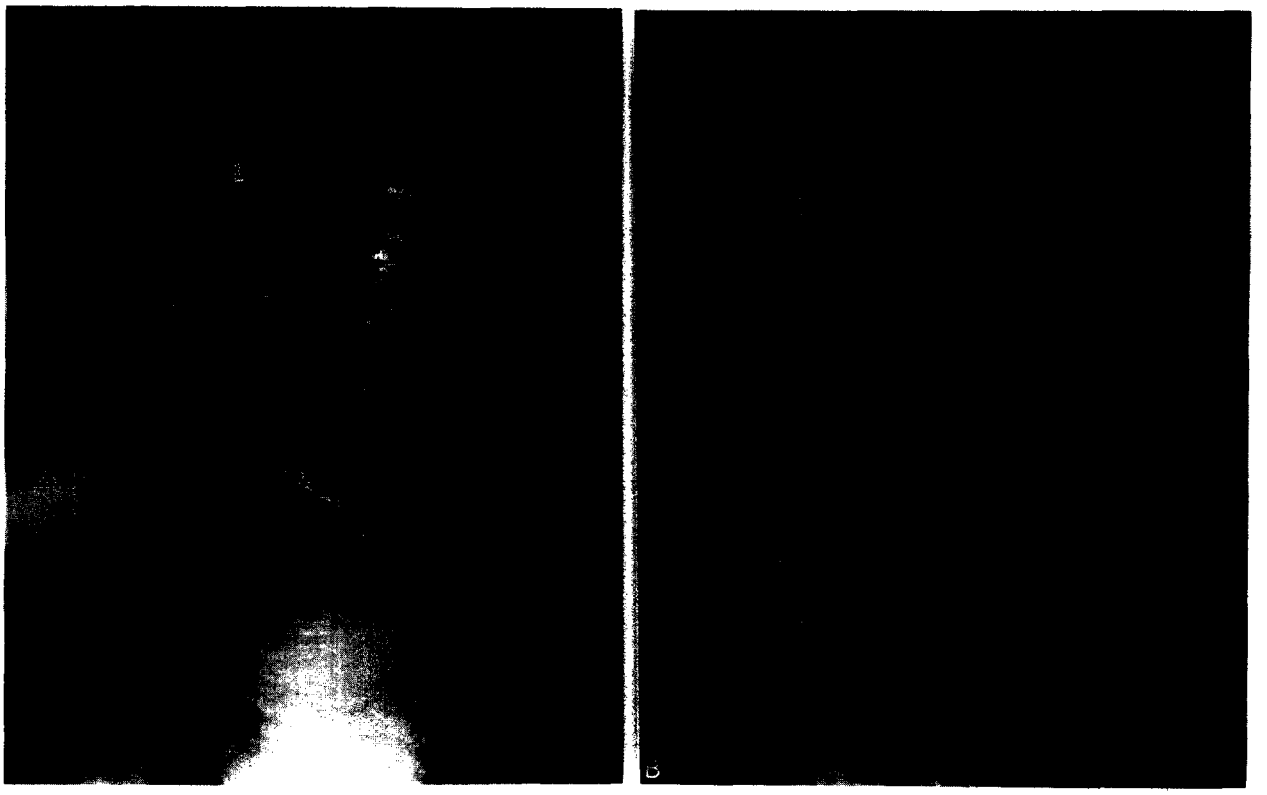

Fig. 3

Figure 3 (A) Dorsum of hand of 8-year-old girl, 3 months after surgery. (B) Palmar view of the same patient.

Anaesthesia

General anaesthesia was performed by mask narcotisation with nitrous oxide and halothane. Trimeperidine hydrochloride and atropine were used as a premedication. During first dressings we used general anaesthesia with ketamine in doses according to body weight.

Operative technique

As the ability of the skin to epithelialise diminishes with age, we recommend carrying out the operation as early as possible. To create the interdigital spaces we incise the epidermis between two fingers to their bases on both sides (palmar and dorsal). Without removing the "epidermal glove", we then separate the fingers using a blunt instrument, without sharp dissection, to the base of the web space. In small children, the web is situated almost at the right place, but in older children the web is still too distal and requires deepening by incision to the appropriate depth. Contractures are released by multiple transverse skin incisions on the palmar side of fingers with subsequent slight extension.

The most important objective is to release the adducted thumb and recreate the first web space by making an incision that extends from the base of the web at the level of the carpometacarpal joint from the dorsum to the palm of the hand. It is important to avoid surgical injury to the neurovascular bundles in the palm. Mechanical extension of adductor muscles must be carried out to allow adequate release of the adduction deformity. Any areas of epidermis that become detached are replaced on the dermis, as in our experience they are able to survive. The wounds are covered with antiseptic dressing.

A plaster of Paris splint is absolutely necessary to immobilise the fingers in a straight position with the thumb in palmar abduction, taking great care to maintain finger separation with dressing material. In advanced hand deformities with total loss of hand function, we do not try to achieve complete extension of fingers during operation and apply a plaster dressing in a flexed position so as not to compromise the blood circulation. During subsequent dressings we straighten them by serial splinting. It is very important to maintain the abducted position of the thumb by careful fixation during each dressing.

Postoperative course and results

Broad spectrum antibiotics are given during the first postoperative week. The first dressing is done on the fifth to seventh day after the operation under general anaesthesia, correcting any finger flexion with a new splint, and an antiseptic dressing is applied. Any bleeding is controlled with diluted adrenaline.

Dressings are carried out every other day thereafter using ointments containing antibiotics and epithelialising components. A general anaesthetic is no longer needed, as the child no longer feels anxious and is able to co-operate. Usually, we use potassium permanganate solution to separate the dressing from the wound.

Complete epithelialisation usually occurs by the third to fourth week after the operation. The parents are cautioned about possible relapse and the importance of following instructions carefully about postoperative splinting and manipulative treatment. Once epithelialisation is well under way, the child may be discharged to have dressings done twice a week. A light plaster volar splint is worn constantly for a month and then worn at night. Gentle elastic pressure dressing is recommended for wear during the day.

In our initial approaches, we separated only one pair of fingers (in six children). They all did quite well, thus encouraging us to operate on all affected fingers on one hand at one operative procedure, operating on the second hand 4–6 months later. We have carried out 49 operations in the last 3 years. Of 15 children with a minimum of 1 year of follow-up, we observed recurrences in eight (53.5%). In all those cases, there was less than perfect postoperative care.

Discussion

There are several reasons for operating without skin grafting:-

1. The skin in young children with RDEB re-epithelialises rapidly.
2. It avoids the need for a skin graft, which is difficult to take because of the tendency for the epidermis to separate as a result of the general skin defect.
3. It is an attempt to simplify surgical treatment, considering the high incidence of recurrences and the need for repeated operations in the future.

We consider surgical treatment only as a first step in rehabilitation of hand function. A rational splint schedule, manipulation and medical treatment are necessary not only to prevent recurrence, but also for growth and development of the hand. In those patients who had operations carried out elsewhere with skin grafting and in our patients who neglected the instructions about postoperative treatment, recurrences occurred about 1–10 months after operation, regardless of operative method. The first sign of recurrence is an adducted position of the thumb, followed by the development of flexion contractures and finally syndactylies.

Most authors consider it is necessary to release not only the skin but also the fascia and intrinsic muscles to overcome the contracture in the thumb.[1,2] As a dermal fibrosis is the main reason for the adducted position of the thumb, incision of the skin with subsequent extension of the muscles is enough to release the thumb, and if during the postoperative phase it is not allowed to return to an adducted position, then the results are excellent.

Scar formation continues well after complete epithelialisation and that is why in the out-patient care, it is very important to check that immobilisation is appropriate. Unfortunately, most of our patients live in small towns and villages far from Moscow, where local surgeons are not well aware of this disease and this makes out-patient care difficult. We are now trying to modify the post-operative approach to avoid rapid recurrence.

Our new approach to the operative treatment of hand deformities in RDEB has several advantages:-

1. Short operative period not exceeding 20 min.
2. Simple operative technique.
3. Avoidance of difficulties connected with skin grafting, including additional trauma. However, it does not diminish the incidence of recurrence, which is high, considering the activity of disease.

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