“The Surgical Management of the Hand Manifestations of Harlequin Ichthyosis”

- 1. Dr Joseph Overland MBBS, GradDipSurgAnat ***
- 2. A/Prof Bruce Johnstone MBBS, FRACS ***

*Department of Plastic and Maxillofacial Surgery, Royal Children’s Hospital, Parkville, Victoria

3052

Corresponding author details:

Dr Joseph Overland
e: overland.joe@gmail.com
m: 0432 351 025
Royal Children’s Hospital
50 Flemington Rd
Parkville, VIC 3052

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A 7-year-old Palestinian female born with Harlequin Ichthyosis was referred for management of severe flexion contractures of the fingers, adduction contractures of both thumbs and pseudosyndactyly causing significant functional impairment. The contractures affected the metacarpophalangeal joints and interphalangeal joints of all fingers and the thumb of both hands. The eyelid deformities were assessed by an ophthalmologist. The patient was also commenced on Acitretin several months prior to the surgical procedure. Retinoid therapy in ichthyosis has been shown to reduce thickness of scaling, increase the ability to sweat and to improve ectropion.¹ Topical therapy included 8 hourly bathing in dilute salt and sodium hypochlorite followed by the application of soft white paraffin ointment. The left hand and upper lid were selected for the first procedure. Infection, uncertainty of graft take in this rare condition, possible poor donor site healing, failure to achieve worthwhile contracture correction and contracture recurrence were particularly discussed with her guardian

A large ellipse of full thickness skin was harvested from the patient's left lower abdomen and was fenestrated. The donor site was closed primarily. The thinnest skin was used by the ophthalmologist as an inlay graft following a generous transverse release of the left upper lid. The first web-space was transversely incised, exposing the underlying subcutaneous tissues. The neurovascular bundles were identified and preserved. Skin graft was fashioned to resurface the 1st web space defect and secured with absorbable sutures.

The 2nd and 3rd web spaces were similarly released. The release was extended over the volar surfaces of the index and middle fingers to correct the contractures. The extent of ulnar sided volar release was limited by the availability of full-thickness graft and poorer potential to correct the more severely contracted and less functionally important little finger. After the defects were fully covered, axial Kirschner wires were placed through the interphalangeal and metacarpophalangeal joints of the thumb, index and middle fingers, holding them in extension. Following thorough lavage of the graft beds, a dressing of tulle gras and silver impregnated hydrogel was applied and secured with gauze, bandages and an above elbow clam shell cast.

As there was minimal postoperative discomfort, she was discharged on day 1 and returned for a dressing change under anaesthesia on day 7. Complete graft take was confirmed, many sutures were trimmed or removed, Kirschner wires were withdrawn and dressings and a new splint were applied. Much of the keratinized superficial epidermis had separated from the adherent vascularized dermis and deeper epidermal elements. At a further dressing change on day 19 in clinic, complete graft take was again confirmed as well as features of very rapid epidermal regeneration. Her carers were then able to perform daily dressing changes and recommence her daily 8 hourly bathing regimen incorporating her left hand.

Similar changes occurred in the upper eyelid graft which had also contracted by about 50% but was still contributing to a very effective reconstruction. As the grafts matured, a moulded silicon elastomer product was applied to protect, compress and conform the healing tissues and was combined with a thermoplastic resting splint to maintain maximal correction of contractures. Progressive functional use of the hand was permitted as the splinting was reduced to nighttime only.

A successful outcome allowed the same procedures to be carried out on the right hand and upper eyelid 3 months after the left sided surgery. There was some pseudomonas infection mainly confined to the dressings at the first change of dressings under a brief anaesthetic at 1 week. This promptly responded to topical treatment including more frequent dressing changes and a faster transition to 8 hourly bathing. By 3 weeks good re-epithelialization was established.

Improved function and appearance has been well maintained 12 months after the initial surgery however a tendency to recurrent contracture has been minimized with ongoing night splinting.

The current body of literature on surgical management of Harlequin Ichthyosis is small and focused largely on limb constriction/encasement^{2 3} or correction of the associated ectropion.^{4 5 6} The rarity of this disease makes its manifestations a challenge for any clinician who encounters it. The severe flexion contractures of the fingers, adduction contractures of the thumbs and pseudosyndactyly in this case were corrected using full thickness skin grafting, with substantial improvement in both function and aesthetics of the hands.

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Author/s:

Overland, J;Johnstone, B

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