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Cochlear implant extrusion in a child with keratitis, ichthyosis and deafness syndrome

S. M. HAMPTON, F.R.C.S.*, J. G. TONER, F.R.C.S.*, J. SMALL, F.R.C.S.†

Abstract
We present a child with keratitis, ichthyosis and deafness (KID) syndrome implanted with a Nucleus device. We discuss the wound complications of this child and the steps taken to deal with the problems encountered when the wound failed to heal, followed by the partial extrusion of her implant. Early surgical management involved resuturing the wound but when this failed a rotational flap was required to cover the implant package and allow eventual healing. Despite the wound problems and revision surgery she has a good audiological result.

Key words: Cochlear implant, complications

Introduction

Keratitis, ichthyosis and deafness (KID) syndrome is a congenital condition encompassing a triad of signs, one of which is profound deafness. As these children also have visual impairment from their progressive keratitis it seems a logical step to offer sufferers cochlear implantation to improve their communication skills. Unfortunately, surgery is not without risk of wound infection, as this case shows, and so each child should be evaluated on an individual basis. We discuss the possible aetiology of the extrusion and outline the surgical measures taken to treat her wound breakdown, culminating in a rotational flap.

Case report

An eight-year-old girl with KID syndrome was referred for cochlear implantation. She had profound bilateral hearing loss, her aided responses to free-field testing were greater than 80 dB in the frequency range 250 Hz to 1000 Hz with no response at higher frequencies. She underwent cochlear implantation which was uneventful. A vertical post-aural incision was used, in which the musculoperiosteum is divided separately 1 cm posterior to the skin incision. This incision is used in all our patients and ensures that the front end of the implant package is well covered. She received a broad spectrum antibiotic for the prophylaxis of infection.

Initially her healing appeared satisfactory, however four weeks post-operatively she developed a wound infection with partial dehiscence. She was promptly returned to theatre and the wound was resutured under general anaesthetic. At the same time the wound was swabbed and the appropriate antibiotic prescribed. Again she appeared to be healing, unfortunately after several weeks she returned with a more extensive wound dehiscence (Figure 1). She then had a formal repair of the implant site carried out.

The necrosing edges of the deficit were excised, with adequate margins, back to healthy skin all around and the posteriorly based rotation flap was marked (Figure 2). The

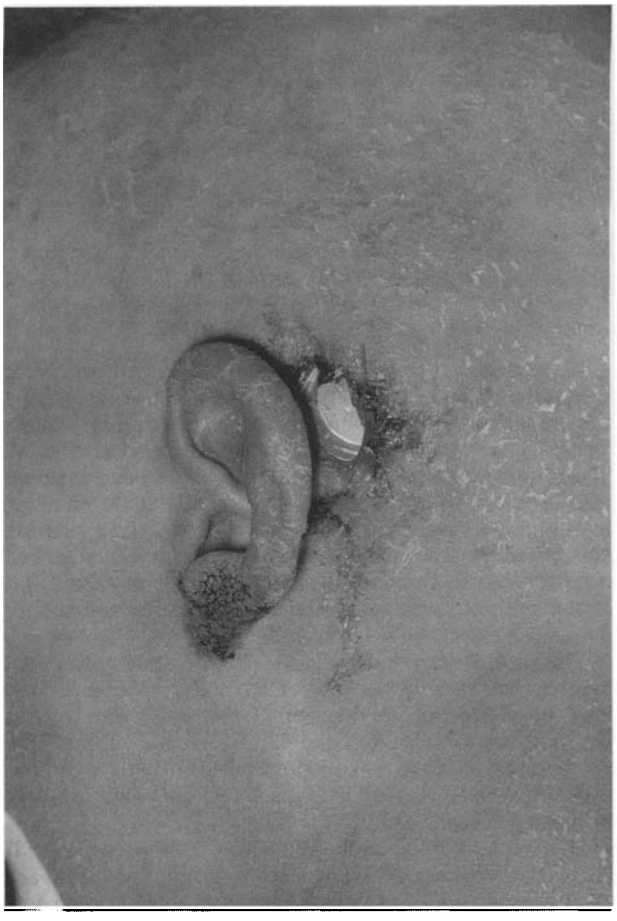

FIG. 1
Extensive wound dehiscence with partial extrusion of the implant.

From the Cochlear Implant Centre, Belfast City Hospital Trust* and the Plastic Surgery Department, Ulster Hospital, Dundonald†.
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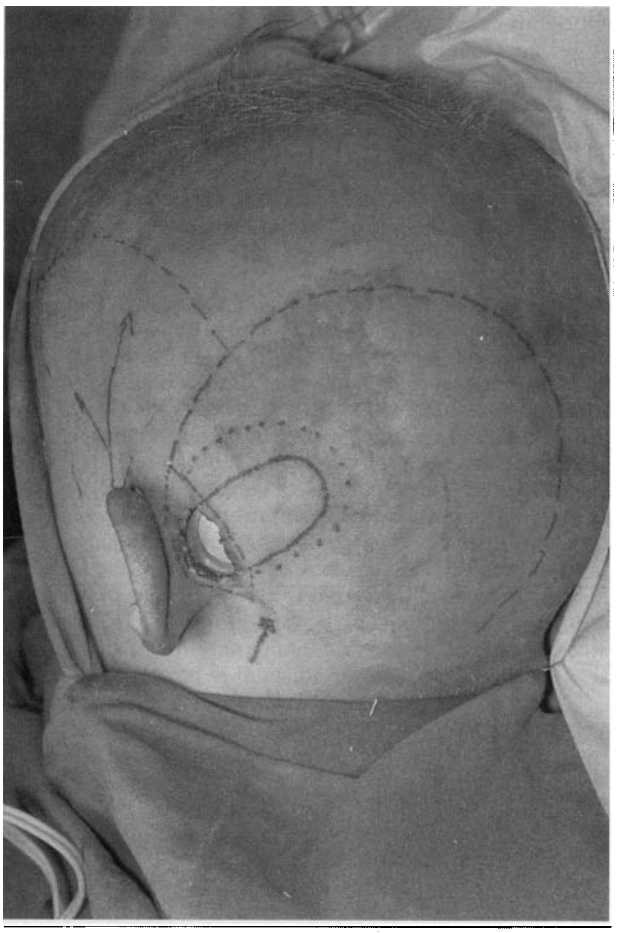

FIG. 2
Skin markings showing the outlines of the implant receiver, the temporalis muscle and fascia flap and the scalp flap.

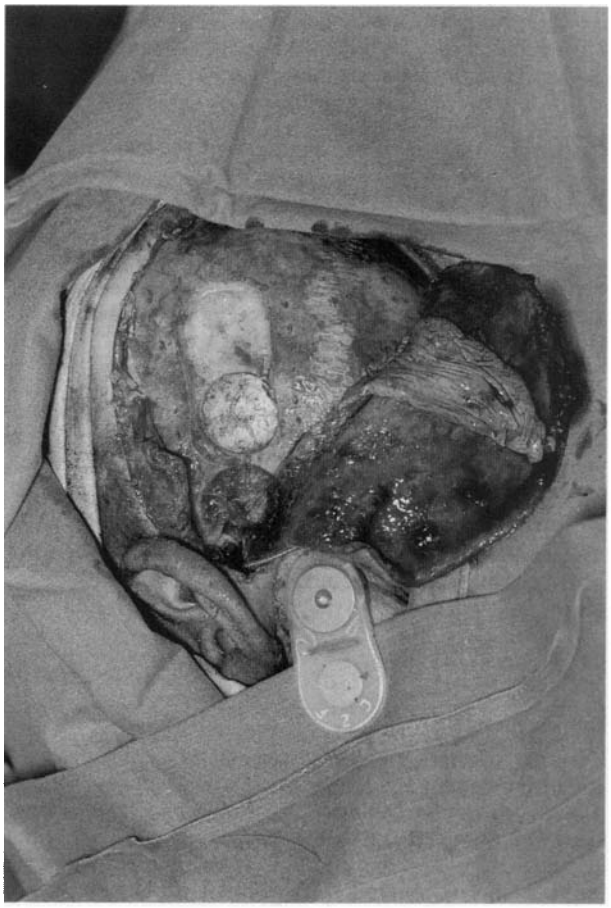

FIG. 3
Intra-operative photograph showing the receiver bed resited posterior to the original site.

implant receiver was removed but the intra-cochlear electrodes were left undisturbed. The mucosa underlying the implant was excised and the receiver bed re-fashioned. The package was replaced and sutured in place with vicryl. The anterior part of the implant which had been exposed was covered with a temporalis muscle and fascia flap, based on the superficial temporal vessels. A scalp rotation flap based on the left occipital vessels was elevated and rotated to cover the post-auricular defect. The wound was closed with absorbable subcutaneous sutures and monofilament interrupted skin sutures.

The sutures were removed after 15 days but subsequent healing was slow, especially at the anterior edge of the implant where the previous breakdown had occurred. It was felt that a revision of the flap was required, this time the implant package was re-sited posteriorly to avoid the problem area (Figure 3). The results of this repair are very promising (Figure 4).

Her implant-aided audiogram show responses of 50 dB across the frequencies. Her speech discrimination skills are improving steadily and she is making good attempts at speech production and imitation.

Discussion

The most common complication associated with cochlear implant surgery involves the skin flap, irrespective of the type of incision used (Cohen *et al.*, 1987; Cohen and Hoffman, 1991; Webb *et al.*, 1991; Hoffman and Cohen, 1993). Wound breakdown can result in exposure of the implant and necessitate the explantation of the device. Both early (Schweitzer and Burtka, 1991) and late (Haberkamp and Schwaber, 1992; El-Naggar and Hawthorne, 1995; Harrison *et al.*, 1995) wound breakdown has been described in the literature, and it is felt that meticulous planning of the incision and avoidance of pressure on the overlying scalp from movement of the device can prevent most of these complications. Despite considerable care being taken, our patient suffered partial extrusion of the implant although excess skin loss and explantation were avoided by prompt surgical intervention.

KID syndrome was named in 1981 by Skinner *et al.* after the three contributing signs, although the common components had been associated as far back as 1915 by Burns *et al.* The aetiology is unknown but the sensorineural deafness has been attributed to cochleosaccular degeneration as part of a general ectodermal dysplasia (Myers *et al.*, 1971; Tsuzuku *et al.*, 1992). Sufferers are particularly prone to cutaneous infections (Baden and Alper, 1977; Skinner *et al.*, 1981; Muramatsu *et al.*, 1987; Hazen *et al.*, 1989; Mallory *et al.*, 1989; Helm *et al.*, 1990) including wound infections, and this is the cause of significant morbidity and mortality.

Our patient had a history of poor wound healing and after a previous Achilles tendon repair had wound breakdown. She also has alopecia associated with the syndrome and wears a wig to conceal this, she refused to do without it on discharge from the ward. Unfortunately the wig has a heavy braid on the inside edge, which is in

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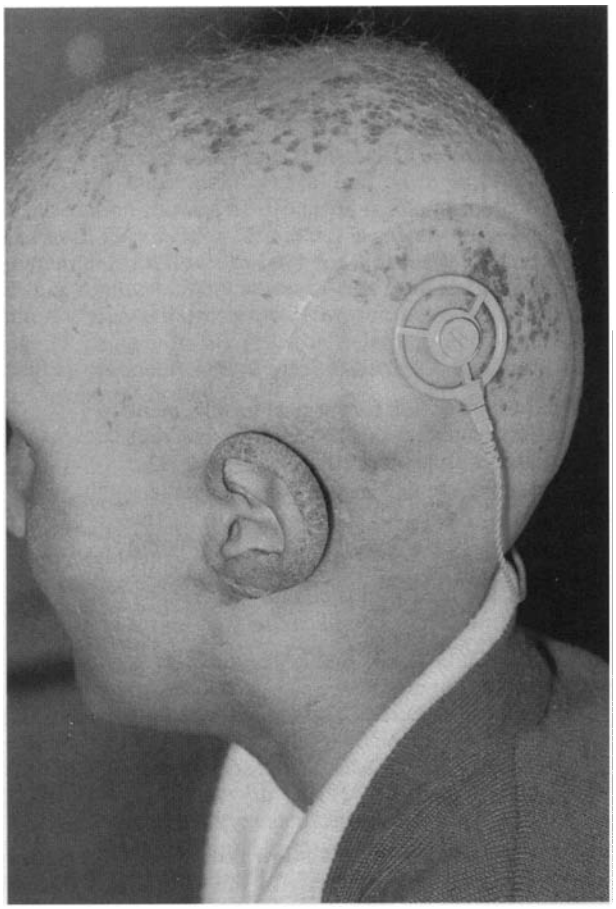

FIG. 4
Flap site six months post-operatively.

contact with the post-auricular area, we felt that the constant rubbing from this coupled with the weight of the v-mic contributed to wound breakdown. Webb *et al.* (1991) discuss a similar case in which they attribute wound breakdown to the pressure of heavy spectacles on the skin overlying the implant.

As we had implanted another child with KID syndrome without any wound problems we looked for any other factors which could have contributed to the wound breakdown and we noted that this girl was on long-term retinoid therapy which the other child was not, an uncommon side-effect of this group of drugs is slow healing.

This case illustrates that KID syndrome does not preclude cochlear implantation, but surgery on affected subjects carries a significant risk of post-operative complications and thus each child should be evaluated on an individual basis.

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Address for correspondence:
Ms S. M. Hampton, F.R.C.S.,
Cochlear Implant Centre,
Belfast City Hospital,
Lisburn Road,
Belfast BT9 7AB.

Fax: 01232 263549.