Patient Information
DEBRA-International's BEST CLINICAL PRACTICE GUIDELINES for skin cancer management in EB

January 2016

DEBRA International has recently published best clinical practice guidelines for the management of squamous cell carcinoma (SCC) in EB patients. These guidelines were developed following a systematic literature review and international expert consensus, which included Australian experts Dedee Murrell and Susan Robertson.

The aim of these guidelines is to best inform health care professionals and patients living with EB on diagnosis and management of SCC to improve patient outcomes and quality of life. You are receiving this information sheet as DEBRA Australia is passionate about informing EB patients and families about the latest available resources and we hope to encourage you to read these guidelines and discuss them with your dermatologist and other treating doctors.

A full copy of the guidelines can be accessed via the link below or the QR code. Please note DEBRA International plans to update these guidelines every 3 years:

http://onlinelibrary.wiley.com/doi/10.1111/bjd.14104/epdf

The main summary findings of the guidelines are listed below. While DEBRA has made its best efforts to prepare this summary, this does not replace the more detailed advice provided in the guidelines and by your treating clinician. It is the individual's responsibility to fully inform themselves further.

- Some subtypes of EB, especially **severe generalized recessive dystrophic EB (RDEB-SG)**, are associated with the development of mucocutaneous SCC.
- These **tumours develop at sites of chronic blistering**, wounds and scarring, behave aggressively and are a leading cause of morbidity and mortality in at-risk EB patients.
- SCC has been reported in patients as young as 6 years of age, and frequently in patients under the age of 20. Cumulative risk of developing SCC increases with age and is parallel to increasing risk of death from SCC.
- **Many EB subtypes, especially EB Simplex, are not associated with increased risk of SCC.**
- Regular skin checks should be performed by professionals with expertise in EB in all at risk groups (consult the guidelines for the list of at risk groups).
- In particular patients with RDEB-SG should have a full skin examination every 3–6 months from age 10 years.
- Clinical screening for lower-risk groups should usually commence from age 20 years and take place every 6–12 months,
- Clinical detection of SCCs in patients with chronic ulcerations is particularly challenging and suspicious lesions should be biopsied for histological evaluation.
- **Ongoing 3-monthly** full skin checks of patients with previously diagnosed SCCs is vital.
- Patients presenting with SCC require a **multidisciplinary review** and SCC lesions >5cm should be imaged, preferably with magnetic resonance imaging.
- **Wide local excision is the treatment of choice,** however amputation of the digit or limb may be required.
- **Radiotherapy may be a useful palliative modality for inoperable EB SCCs.**
- Conventional chemotherapy may be of some benefit but risks may outweigh the benefits.
- Limb prostheses may be used successfully in EB following limb amputation.
- **Psychological support of the patient with EB and family/carers is vital after a diagnosis of SCC and as end of life care approaches.**

Reference: Mellerio J.E. et al., (2015) "Management of cutaneous squamous cell carcinoma in patients with epidermolysis bullosa: best clinical practice guidelines". *British Journal of Dermatology* DOI 10.1111/bjd.14104.