



Guidelines of Care of Epidermolysis Bullosa (EB) Cases As'ad Al-Hamad Dermatology Center, Kuwait

Diagnosis of a EB Case:

A detailed clinical history, examination, and investigations are recorded for each case registered as EB in our Sick-kids clinic

1. Clinical Data:

- a. Birth history/antenatal history of mother
- b. First appearance of blisters
- c. Family history/pedigree/presence or absence of consanguinity
- d. Presence/absence of scarring/milia/pigmentary abnormalities
- e. Associated abnormalities: Mucosae/nails/teeth/hair/palms & soles/systemic
- f. Photosensitivity
- g. Sequelae
- h. Developmental parameters
- i. Appearance of new abnormal skin lesions

2. Skin Biopsy:

- a. Electron Microscopy
- b. Immunofluorescence/immunohistochemical antigen mapping

3. Routine Investigations:

- a. Complete blood counts
- b. Serum biochemistry
- c. Skin swabs: culture & sensitivity
- d. Urinalysis
- e. Others as indicated

4. Mutational Screening*: for responsible gene



Final Diagnosis: (Classification adapted from – *Fine J-D et al. J Am Acad Dermatol 2008; 58: 931-50*)

- a. EB-Simplex
 - Suprabasal
 - Basal
- b. EB-Junctional
 - Herlitz
 - Others
- c. EB-Dystrophic
 - Dominant
 - Recessive
- d. Kindler's Syndrome

Management: Treatment needs to be tailored according to EB subtype

a. Skin care:

- i. Avoidance of friction/trauma
- ii. Gentle skin handling
- iii. Appropriate clothing
- iv. Wound Care:
 - Antiseptic cleaning
 - Specialized wound dressings: Mepital/Mepiplex

b. Antibiotics: Periodic use of topical/systemic antibiotics as indicated

c. Nutritional supplements:

- a. High protein diet
- b. Proteins/multivitamins/iron/calcium/magnesium/zinc supplements as indicated

d. Multidisciplinary Approach: Patient's referral are arranged to different sub-specialties as indicated:

- a. Dentist
- b. Gastroenterologist
- c. Plastic surgeon
- d. Pediatrician
- e. Dietician
- f. Others as indicated

e. Genetic Counseling: Based on type of EB after the final diagnosis has been reached

*** Currently, the facilities for mutational screening are not available in Kuwait so mutational screening of our patients with EB was carried out in National EB Lab. Guy's hospital, London**