

Epidermolysis bullosa (EB)-Insel :

Towards a national competence center for the diagnosis and management of EB patients

C. Gouveia¹, S. Häfliger¹, R. De Lorenzo¹, K. Schwab², K. Neuhaus³, S. Schibli⁴, S. Berger⁵, P. Folly², T. Zürcher², G. Steinmann², A. Dupenthaler⁶, F. Stucki⁷, S. Galatti⁸, E. Vögelin⁹, S Eychmüller¹⁰, E Seifert¹¹, A. Schwieger-Briel¹², P. Itin¹³, E. Gianadda¹⁵, L. Borradori¹; C. Aebi²; K.Kernland-Lang^{1,14}

1. Department of Dermatology, University Hospital of Bern, Bern | 2. Department of Pediatrics, University Hospital of Bern, Bern | 3. School of Dental Medicine, Department of Preventive, Restorative and Pediatric Dentistry, University of Bern | 4. Division of Gastroenterology, University Children's Hospital of Bern, Bern | 5. Department of Pediatric Surgery, University Hospital of Bern, Bern | 6. Division of Pediatric Infectious Diseases, University Hospital of Bern, Bern | 7. Department of Anesthesia and Pain Medicine, University Hospital of Bern, Bern | 8. Division of Human Genetics, University Children's Hospital of Bern, Bern | 9. Department of Orthopedic, Plastic and Hand Surgery, University Hospital of Bern, Bern | 10. University Center for Palliative Care, Inselspital, University Hospital Bern, Bern | 11. Division of Phoniatrics, Department of Otorhinolaryngology, Head Neck Surgery, University of Bern, Bern | 12. Department of Pediatric Dermatology, University Children's Hospital, Zurich | 13. Department of Dermatology, University Hospital of Basel, Basel | 14. Department of Pediatrics, Hospital of Baden, Baden | 15. Fondation Enfants Papillons, Sion | Switzerland

Hereditary EB is a rare group of mechano-bullous disorders characterized by fragility and blistering of the skin and other specialized epithelia, such as mucosae. EB has a devastating impact on the life of the patient and the caregiver. According to Dystrophic Epidermolysis Bullosa Research Association (DEBRA), 1 out of 50,000 babies born are affected with EB.

In Switzerland, the estimated number of patients with moderate and severe EB forms is between 110 and 130. So far, there is no curative treatment EB. An A multi-disciplinary specialized team is required to ensure proper management of EB patients. Specialized supportive wound care and early treatment of complications are essential.

In the past 10 years, we have created in Bern a multidisciplinary, integrative EB center. It is based on an unique patient-centered concept utilizing the specific expertise of several health professionals important for a comprehensive patient's management, involving specialized nurses, gastroenterologists, physiotherapists, dieticians, social workers, pediatricians, anesthetists, hand and pediatric surgeons, infectiologists, ENT specialists, oncologists, and dentists.



Fig. 1, 2 Hand surgery for pseudo-syndactyly correction and occupational therapy.



Fig. 3, 4. Blistering in Esophagus. Fig 5. Gastrostomy after esophageal stenosis



Fig. 6,7 Aggressive squamous cell carcinoma; patient starting Nivolumab therapy.



Fig.8 Extensive erosions



Fig. 10 Pain therapy



Fig.9 Microstomia; caries



Fig. 11 After a successful home visitation

We provide information and support to affected people and family members and interact with all relevant institutions (home care teams, schools, professional and patients associations...). Our team is completed by our EB-nurse expert that supports and counsels the patients at home and acts as liaison person to bridge the gap between hospital, primary care, home and school.

We collaborate with other Swiss Hospitals (such as USB and KISPI-UZH), EB international clinical and research centers. The competence center is generously supported by private institutions, the contribution of which is critical to ensure the services provided.