



**6th Genodermatoses in Mediterranean working session  
1st TAG meeting**

May 22 - May 25, 2009, Greece

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# Epidemiology of genodermatoses

- Slovenia has a population of 2 millions people.
- Darier's disease – 54 registered patients
- sporadic cases of:
  - severe congenital ichthyoses
  - Erythrodermia ichthyosiformis congenita
  - Neurofibromatosis
- The exact number of patients with other genodermatoses (palmoplantar keratoderma, ichthyosis vulgaris and others) is not known, because patients with mild disease do not seek dermatological treatment

# Epidermolysis bullosa - Prevalence in Slovenia

- EB dystrophica: 5 patients (3 children)
- EB junctionalis: 2 patients
- EB simplex: 20 children in 4 families;  
the exact number of adult patients is not  
known; in some of them disease is  
mildly expressed

# Needs of the patients-children

- All children with genodermatoses are treated at the Department of Dermatovenereology in Ljubljana
- Cooperation with paediatricians and other specialists is provided
- Education of parents about the needs and care of their children
- Cooperation with social workers is well established

## Needs of the patients – children with Epidermolysis bullosa

- regular dermatological examinations of children at least three times per year
- hospitalization of children at least once per year for detailed examinations
- regular bandaging of skin wounds is provided at home by experienced nurses
- since 1993, in all children diagnosis of EB was confirmed histological and with electron microscopy

# Needs of adult patients

- all adult patients with Epidermolysis bullosa are treated at the central Department of Dermatovenereology in Ljubljana
- all patients with dystrophic and junctional BE are hospitalised once a year for detailed examination of the whole skin and other examinations
- patients with other genodermatoses are treated at different dermatological centres
- for severe patients skin care by home nursing is provided
- costs for the management of patients are assured by the Insurance company with restrictions regarding certain bandages, creams and topical treatment

# Dermatological centres in Slovenia

- In Slovenia there are two University Departments of Dermatovenereology and two additional hospital centres.
- proceeding of patients according to multidisciplinary approach is provided in all dermatological centres

# Slovenian DEBRA mission

- foundation in 2005
- units patients with Bullous Epidermolysis and their relatives, changing experiences about the disease
- support to patients and their families
- to provide help in socializing the patients
- education and information about new achievements regarding skin care and treatment for patients, relatives and professionals
- assistance to provide new wound dressings
- it helps in qualifying professional teams



# Debra Slovenia

- efforts for public information of the disease
- collection of additional financial resources to support the patient's needs

Koža, krhka in ranljiva kot krila metulja ..., zabodeni pogledi, polni pomilovanja, včasih celo zgražanja ..., na videz drugačnost, v svojem bistvu pa enake želje in pričakovanja, kot jih imajo ostali, »normalni« ljudje ..., vsak nov dan s svojim prihodom prinese tudi nove rane, oskrbo le-teh, nove zabodene poglede, vedno novo bolečino, boleče zdravniške posege, vendar je ni čez srčno bolečino, katero povzroča stalna odvisnost od drugih ..., vse to in še več se skriva za »samo« dvema besedama:

## Bulozna Epidermoliza.



## ... v razmislek

Skrb za sočloveka je v domeni vseh in **vsakdo** je bil prepričan, da bo za to poskrbel **nekdo**.

Lahko bi naredil **kdorkoli**, a ni naredil **nihče**.

**Nekdo** je ostal brez pomoči, katero bi lahko ponudil **kdorkoli**, vendar **nihče** se ni zavedal, da je **vsakdo** ne bo ponudil.

Konča se torej tako, da **vsakdo** prevali odgovornost na **nekoga**, medtem ko **nihče** ne naredi tistega, kar bi lahko **kdorkoli**. Torej...

Bodite tisti **NEKDO**, ki pomaga lajšati bolečino.

Kako?

Delo društva je odvisno od vašega razumevanja in podpore:

TRR: 03131-1000224363

SKB BANKA D.D., LJUBLJANA

Lahko pa se tudi včlanite v društvo in s svojimi idejami in sposobnostmi pripomorete k delovanju društva. Za dodatne informacije smo vam na voljo na spodaj navedenih podatkih:

DRUŠTVO DEBRA SLOVENIJA

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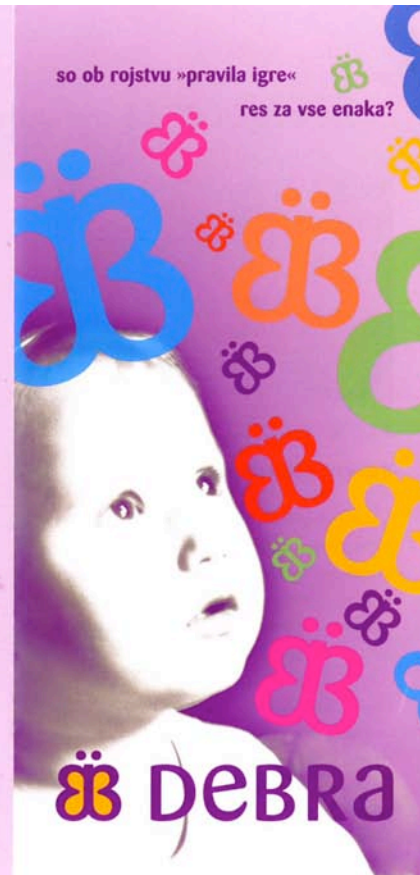
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George Banka Mednarodna

so ob rojstvu »pravila igre«  
res za vse enaka?



# Multidisciplinary professional team of experts for patients with Bullous epidermolysis

- professional team of different specialists, needed in the care of children and adult patients with EB was established in 2009
- cooperation between dermatologists, paediatrics, genetics, specialists for ophthalmology, dental medicine, plastic surgery, internal medicine, psychology
- to improve the knowledge about EB among different specialists and to improve treatment possibilities for patients

# Treatment of patients with genodermatoses

- regularly check-ups of patients at dermatologists in patients with severe genodermatoses
- multidisciplinary medical care
- treatment is directed towards state-of-the-art recommendations and possibilities

# Collaboration with European centres

- cooperation between Debra Slovenia and Debra Croatia and organisation of some meetings in the last years
- contacts with Department of Dermatology, Molecular Dermatology, in Germany (Freiburg) were established with the intention of performing genetic analysis in our patients with dystrophic and junctional Epidermolysis bullosa

# Education

- participation of two dermatologists, two nurses, and one paediatrician, at the Symposium on Epidermolysis bullosa in UK and Croatia in 2009

# Future plans

- to prepare the National Register for all genodermatoses in Slovenia
- to organise national network for genodermatoses in already existed dermatological centres
- to improve collaboration on the European level
- to improve diagnosis and treatment
- further education of younger dermatologists and other caregivers about genodermatoses