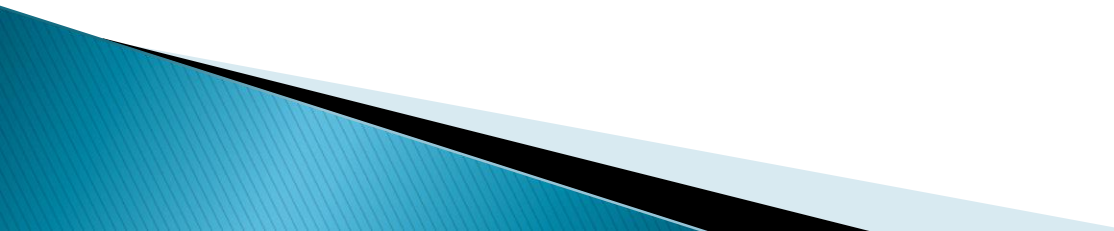


# Quality of life of patients with epidermolysis bullosa

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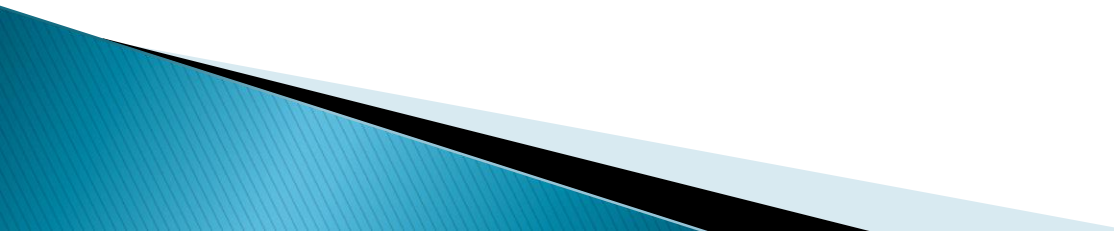
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- ▶ Having a vulnerable skin causes many limitations in everyday life and numerous psychological and sociological problems for the patients and their families
  - ▶ There is a growing need to investigate the patients perspective of the impact of the disease on their lives
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# HRQoL

- ▶ health-related **quality of life** (HRQoL): is an individual perception of how her/his wellbeing is affected over time by a disease, disability, or a disorder and how it affects emotional, social and physical, cognitive, work-related aspects of the individual's life
- ▶ Determination of HRQoL gives information of an individual's physical, emotional health, psychological, social wellbeing, economic burden, fulfilment of ones personal expectations, and functional capacity to carry out daily routines

# Studies of QoL in EB

- ▶ Published studies are scarce
  - ▶ usually involve a relatively small number of patients
  - ▶ variable disease severity
  - ▶ studies are: often descriptive, use different tools
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# Evaluation of HR QoL

- ▶ **different questionnaires/tools**
  - ✓ Generic instruments
  - ✓ Skin-specific questionnaires: DLQI, Skindex-29 and 17 ...
  - ✓ Disease specific QoL instruments
- ▶ **Studies are often descriptive (interview, semi structured interview)**
- ▶ **Some studies investigate psychological, functional health status, economical effects of the disease and/or the effect on family**

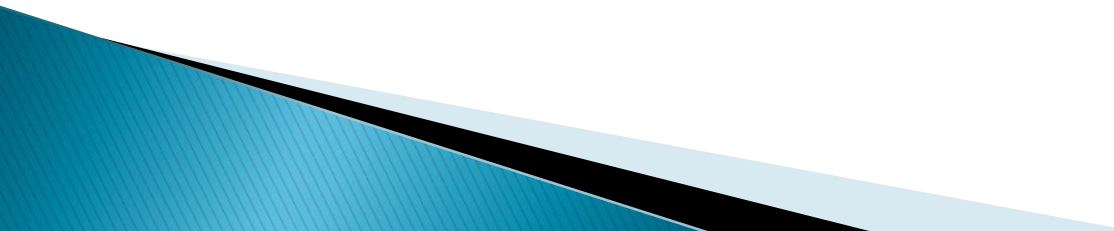
# QoL for EB questionnaires

- ▶ Quality of life evaluation questionnaire in EB (QOLEB)- Br J Dermatol 2009;161:1323-30. (translated and validated to Dutch, Brazilian Portuguese, Hispanic)
- ▶ EB family/parental burden score questionnaire ( EB-BoD)- Br J Dermatol 2015; 173:1405-10.

# Evaluation of HR QoL

- ▶ **different questionnaires/tools**
  - ✓ Generic instruments: SF-36, VAS....
  - ✓ Skin-specific questionnaires: DLQI, Skindex-29 and 17 ...
  - ✓ Disease specific QoL instruments
- ▶ **Studies are often descriptive (interview, semi structured interview)**
- ▶ **Some studies investigate psychological, functional health status (mobility, hand function), economical effects of the disease or the effect on family**

# What have we learned

- ▶ Similar issues as other chronic conditions—coping with symptom management, disability, emotional impact, need for easy access to good medical care
  - ▶ Similar issues as other skin diseases—visible and has an influence on social and interpersonal relations
  - ▶ Specific for EB—is rare, life long and hereditary nature, often disfiguring and painful
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# QoL of adult/children with EB

- ▶ QoL comparable to moderate and severe psoriasis and AD
- ▶ The perceived disease severity and skin area involved are relevant for QoL in all EB subtypes
- ▶ Greatest scores RDEB–HS
- ▶ QoL is more severely impaired in children and women

Clin Experimental Dermatol 2002;27:707–10.  
Clin Experimental Dermatol 2004;29:122–7.  
Br J Dermatol 2009;161:869–77.  
Ann Dermatol 2016;28:6–14.

# QoL of adult/children with EB

- ▶ Very few patients are free of pain, it varies depending on extent of skin surface involved and depth of wounds
- ▶ patients with all subtypes of EB are likely, to at least partially, be dependant on others
- ▶ restriction of physical, social and sporting activities which influences QoL
- ▶ impact of the disease on employment and education

Clin Experimental Dermatol 2002;27:707-10.

Clin Experimental Dermatol 2004;29:122-7.

Br J Dermatol 2009;161:869-77.

Ann Dermatol 2016;28:6-14.

# Impact of EB on psychological distress (depression/anxiety)

- ▶ the physical burden placed on EB patients can lead to depression/anxiety
- ▶ psychological suffering and severity of the disease did not correlate in all studies
- ▶ Possible explanation: patients get accustomed to living uncomplainingly with the disease and value areas of life which remain uncompromised by EB (school, family, friends)

Br J Dermatol 2009;161:869–77.

J Clin Psychol Med Settings 2010;17:333–9.

J Dermatol 2016;43:70–8.

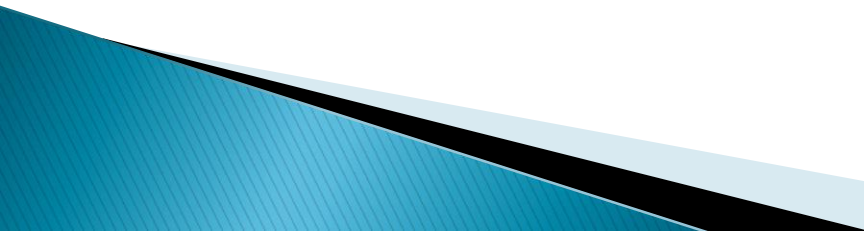
# Patients' perspective

- ▶ importance of having comprehensive information about the disease, the exact diagnosis, mode of inheritance
- ▶ want to be „in charge” of everyday life
- ▶ want to be acknowledged as “partners” in the care process by health care professionals
- ▶ problem of interaction with health care professionals who are not well informed about EB
- ▶ need to interact with those who understand them – other people with EB

# Childrens' perception

- ▶ Itch (new wounds, feeling of helplessness, impact on sleep)
- ▶ constant pain and fear of pain
- ▶ difficulties to join in activities with others due to physical limitations
- ▶ feelings of being left out and loss of self-esteem
- ▶ Stigmatisation – often bothered by the visibility of the disease (annoyed by staring and teasing of others)
- ▶ the feeling of being different – wish to be seen as „normal“ and to have an identity unrelated to their skin disease

# Implications for support provision

- ▶ Informational needs (patients, family, layman, health care providers)
  - ▶ Need for extensive practical, social and emotional/psychological support
  - ▶ Availability of multidisciplinary, specialized medical centres
  - ▶ Need for assistance of parents in seeking support and counselling to prevent destruction of the family unit
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# Conclusions and aims

- ▶ Investigation of the HRQoL is important in care and management of patients with EB – holistic approach
  - ▶ Can help in assessing new therapies in clinical trials
  - ▶ Determination of HRQoL and comparison with other diseases can help in fundraising
  
  - ▶ Aims
    - enrolment of a greater number of patients, with clinically different disease severity
    - in quantitative studies use disease specific questionnaires
    - development of paediatric QoL tools
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