Burden of itch in EB, a cross sectional study

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• What is known?
• Aim of the study
• Methods
• Results
• Pathophysiology of itch in EB?
• Treatment (now and future)
Itch is an unpleasant feeling leading to scratching.

Acute Itch -> scratching -> elimination of hazard

Chronic itch : scratching overschoots it’s aim
Itch in EB

• Common complaint
• Few data in the literature
• EB pruriginosa: subtype of EB characterised rather with prurigo lesions than blisters and bullae
• One study: main complaints in children with EB
  – Itch: most important, before pain !!!

Acta Derm Venereol 2008; 88: 143-150
• Inleiding
• **Aim of the study**
• Method
• Resultats
• Hoe ontstaat jeuk?
• Wat kunnen we er aan doen?
• Toekomst
Analysis of prevalence and characteristics of itch in the 3 major forms of EB: EBS, JEB and DEB
• Introduction
• Aim of our study
• Method
• Results
• Pathophysiology of itch?
• Treatment now and future
– Cross sectional study

– **Tool**: Leuven itch scale: [www.itchscale.eu](http://www.itchscale.eu)

  • 11 questions to evaluate all aspects of itch
  • for patients > 14 years
  • validated in different patient groups (*Burns 2011; 37: 939-950*)
  • translated in different languages and already used in other studies
  • For this study: language was adapted for the Netherlands

– **Sent to the patients and filled out at home**

The measurement of itch: Developent and validation of the Leuven Itch Scale

*Burns 2011; 37: 939-950*
Patient recruitment:
- University Hospital Leuven (Belgium)
- University Medical Center Groningen (the Netherlands)
- members of DEBRA Belgium

Inclusion criteria:
- Diagnosis of one of the three subtypes of EB
- Dutch is native language
- older than 18 years
- provided written informed consent.
What is measured with the LIS?

1. Frequency: How often did you experience itch in the past month?
2. Duration: In the past month, how long, on average did your itching episode last?
3. Timing of itch: In the past month, when did the itching occur?
4. Circumstances: In the past month in what circumstances did the itching occur?
5. Severity: In the past month, how bad was the itching you have been experiencing? (on a VAS scale)
What is measured with the LIS?

6. **Management**: In the past month, how was your itching treated?

7. **Efficacy of treatment**: How satisfied are you with the treatment for your itching?

8. **Consequences**: In the past month, what were the consequences of your itching?

9. **Sensory perception**: In the past month, how did your itching manifest itself?

10. **Distress**: In the past month, how distressing was your itching? (VAS)

11. **Location**: In the past month, which parts of our body itches? On human picture

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Method

- Data are statistically analysed with SPSS 15 for windows
- The findings are represented in a diagram
• Introduction
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20 Belgian 20 Dutch patients were recruited

- Epidermolysis bullosa simplex (EBS) (n = 19)
- Junctional epidermolysis bullosa (JEB) (n = 7)
- Dystrophic epidermolysis bullosa (DEB) (n = 14)
Results: incidence of itch

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Comparison with atopic dermatitis
Circumstances in which itch occurs

<table>
<thead>
<tr>
<th>Circumstances</th>
<th>Total</th>
<th>EBS</th>
<th>JEB</th>
<th>DEB</th>
</tr>
</thead>
<tbody>
<tr>
<td>During change in weather</td>
<td>47%</td>
<td>21%</td>
<td>29%</td>
<td>85%</td>
</tr>
<tr>
<td>During spells of pain</td>
<td>38%</td>
<td>21%</td>
<td>57%</td>
<td>46%</td>
</tr>
<tr>
<td>When making movements</td>
<td>44%</td>
<td>64%</td>
<td>0%</td>
<td>46%</td>
</tr>
<tr>
<td>When sweating</td>
<td>62%</td>
<td>43%</td>
<td>57%</td>
<td>85%</td>
</tr>
<tr>
<td>In a hot environment</td>
<td>65%</td>
<td>43%</td>
<td>86%</td>
<td>77%</td>
</tr>
<tr>
<td>When standing up after sitting or laying</td>
<td>18%</td>
<td>21%</td>
<td>14%</td>
<td>15%</td>
</tr>
<tr>
<td>When stressed</td>
<td>47%</td>
<td>29%</td>
<td>57%</td>
<td>61%</td>
</tr>
<tr>
<td>With contact with air</td>
<td>18%</td>
<td>14%</td>
<td>0%</td>
<td>31%</td>
</tr>
<tr>
<td>When touching the skin</td>
<td>35%</td>
<td>50%</td>
<td>29%</td>
<td>23%</td>
</tr>
</tbody>
</table>

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## Consequences of itch

<table>
<thead>
<tr>
<th>Condition</th>
<th>total</th>
<th>EBS</th>
<th>JEB</th>
<th>DEB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lesions from scratching</td>
<td>85%</td>
<td>71%</td>
<td>100%</td>
<td>92%</td>
</tr>
<tr>
<td>Reduced social contact</td>
<td>38%</td>
<td>29%</td>
<td>57%</td>
<td>38%</td>
</tr>
<tr>
<td>Reduced quality of life</td>
<td>50%</td>
<td>36%</td>
<td>57%</td>
<td>61%</td>
</tr>
<tr>
<td>Disturbance of routine activities</td>
<td>56%</td>
<td>43%</td>
<td>57%</td>
<td>61%</td>
</tr>
<tr>
<td>Difficulties in falling asleep</td>
<td>88%</td>
<td>71%</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>Waking up</td>
<td>73%</td>
<td>71%</td>
<td>85%</td>
<td>69%</td>
</tr>
<tr>
<td>Need for sleeping pills</td>
<td>35%</td>
<td>14%</td>
<td>43%</td>
<td>54%</td>
</tr>
<tr>
<td>Loss of appetite</td>
<td>18%</td>
<td>7%</td>
<td>29%</td>
<td>23%</td>
</tr>
<tr>
<td>Bad mood</td>
<td>53%</td>
<td>43%</td>
<td>57%</td>
<td>61%</td>
</tr>
<tr>
<td>Changes in behaviour towards others</td>
<td>41%</td>
<td>28%</td>
<td>43%</td>
<td>54%</td>
</tr>
<tr>
<td>Loss of concentration</td>
<td>62%</td>
<td>43%</td>
<td>71%</td>
<td>77%</td>
</tr>
</tbody>
</table>
# Sensory characteristics of itch

<table>
<thead>
<tr>
<th></th>
<th>total</th>
<th>EBS</th>
<th>JEB</th>
<th>DEB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tickling (crawling over my skin)</td>
<td>56%</td>
<td>43%</td>
<td>58%</td>
<td>70%</td>
</tr>
<tr>
<td>Tingling (heating up after cold)</td>
<td>21%</td>
<td>7%</td>
<td>14%</td>
<td>38%</td>
</tr>
<tr>
<td>Prickling (being pricked)</td>
<td>38%</td>
<td>21%</td>
<td>29%</td>
<td>61%</td>
</tr>
<tr>
<td>Stinging (piercing skin)</td>
<td>29%</td>
<td>14%</td>
<td>14%</td>
<td>54%</td>
</tr>
<tr>
<td>Burning (in fire)</td>
<td>44%</td>
<td>36%</td>
<td>29%</td>
<td>61%</td>
</tr>
</tbody>
</table>
Conclusions:

• Itch is a frequent problem in EB, more problematic in more severe forms in which it is comparable to itch in AD

• Itch is perceived as more distressing in EB compared with AD, but has less consequences

• Main consequences are lesions due to scratching, sleep disturbances and impairment of quality of life
Future research

• Correlation with the severity and subtypes of EB
  – Birmingham EB severity scale *BJD 2009 : 160 : 1057*

• Research on the mechanism provoking itch in EB

• Research to find better treatments
• Introduction
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• Treatment now and future
Itch in EB

Dry skin

Wound healing and microbial infection

Psychological factors

Internal organ dysfunction

Combination with another skin disease is possible!
Chronic itch

Central sensibilisation

Hyperknesis
Same trigger provokes more intense itch at places which already itch <-> those that don’t itch

Alloknesis
Trigger that usually does not provoke itch, provokes itch (eg contact with the skin)
• What is known?
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• Possible treatments (now and future)?
Prevention of itching and scratching!

- Avoid triggers
  - Avoid heat, wool, use relaxation
  - Avoid deshydration of skin: no powders, no soap or detergents, use emollients

- Alternatives
Medical treatment : add on!

1. Cyclosporine
2. Antidepressants with itch relieving properties (SSRI, TCA, TTCA)
3. Gabapentin or pregabalin
4. Leucotriene receptor antagonist, topical corticosteroid, topical calcineurine inhibitor, doxycyclin
5. Sedative antihistaminics
6. Non sedative antihistaminics

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Other possibilities

• Thalidomid (also report in EB)
• Diaminodiphenylsulfone (report in EB)
• Systemic corticosteroids (but consider side effects)
Future?

Autotaxine inhibitors
LPA-receptor blockers

Anti -VEGF

Anti IL31
H4 blockers

Hstamine lib
Substance P
IL31
Staph

PAR inhibitors
Endo/exogeen

Protease inhibitors

TLR7 <- virussen

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Future?