The psychological adjustment of children with Epidermolysis Bullosa

By

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EB and Children

- Lansdowne, Atherton, Dale, Sproston & Lloyd (1986)
  - Asked parents about psychological and practical difficulties.
  - 16% reported social problems
  - 33% reported worries that their child would have future social problems

- Lansdowne & Nabarro (1990)
  - No psychological differences in children with EB (n=5)

- Titman (unpublished, 2001)
  - No consistent psychological differences (n=28)
EB and Children

- **Horn & Tidman (2002)**
  - Measured dermatology-related quality of life in adults and children.
  - Children with EBS and DDEB had approximately the same amount of impairment as children with severe eczema or psoriasis.
  - Children with RDEB (Hallopeau-Siemens DEB) had far worse quality of life.
EB and Children

- Andreoli, Mozzetta, Angelo, Paradisi & Foglio Bonda (2002)
  - Used the Rorschach projective test on children and adults (N=20)
  - Normal emotional and social development
  - Normal rates of frank psychopathology
  - Positive acceptance of EB, awareness of the realities of EB
  - In some areas, above the norm.
Purpose

- Increase our understanding of EB and its effects
- Identify psychological priorities
- Identify physical aspects of EB that affect psychological functioning the most
- Identify any other factors that can protect children psychologically
Study 1

- GOS patients between 7 and 14
- N=84 (>70% of population)
  - 21 DDEB
  - 18 RDEB
  - 30 EBS
  - 12 No/other diagnosis

- Completed questionnaires in the waiting room when attending routine hospital review at GOS.
Question 1
What is the psychological profile of children with EB?

We asked the children:
1. Anxiety
2. Depression
3. Self-esteem
4. Anger
5. Behaviour
6. Dermatology-related quality of life

We asked the parents:
1. Emotional problems (anxiety/depression)
2. Behaviour
3. Hyperactivity
4. Social skills
5. Peer relationship problems
Question 1
What is the psychological profile of children with EB?

The children said:

- normal overall group means in all areas
- more children scored in the clinical range for anxiety and low self-concept than in the general population
- fewer children scored in the clinical ranges for depression, anger and disruptive behaviour
Question 1
What is the psychological profile of children with EB?

The children said:

- Same about their educational and athletic ability, physical appearance
- Better about their behaviour
- Worse about their social acceptance and self worth
- Better dermatological quality of life than the children with EB in Horn & Tidman (2002) study
Question 1
What is the psychological profile of children with EB?

The parents said:

- group means in the normal range
- more children in the clinical range for emotional difficulties and peer problems than the general community
- same rates of conduct and hyperactivity problems
- more children with above average pro-social behaviour than in the non-EB population
Question 2
What aspects of EB are related to psychological functioning?

We looked at:
- Percentage of skin unhealed
- Gastro-intestinal problems
- Mobility
- Pain
Question 2
What aspects of EB are related to psychological functioning?

Pain was associated with

- quality of life
- self-esteem
- anxiety
- depression
- emotional problems
Question 2
What aspects of EB are related to psychological functioning?

% skin unhealed was associated with:

maternal stress
Question 2
What aspects of EB are related to psychological functioning?

Gastro-intestinal compromise was associated with:

dermatological quality of life
Question 3
Are there any other psychosocial factors that protect/predispose...?

- Overall self-esteem was protective
- Passive coping style made things worse
Other findings...

- Pain and nutritional compromise highly correlated

- Pain and % skin unhealed were not correlated
Differences between EB types

Children with RDEB

- had significantly more physical problems:
  nutritional compromise, unhealed skin, pain and lower athletic ability
- BUT reported few psychological differences:
  better behaviour, less angry, lower satisfaction with appearance
- AND reported using more active coping strategies
Conclusions

- Psychologically well, but more likely to have high anxiety, low self-esteem and unsatisfactory peer relationships.

- Pain is linked to psychological adjustment

- The role of gastro-intestinal pain

- High self-esteem was helpful in protecting the children psychologically

- Passive coping style might exacerbate psychological difficulties

- Very few psychological differences between EB types
The Study(2)

- **We interviewed**
  - EBS (n=11)
  - RDEB (n=4)
  - DDEB (n=3)
  - 8-14 years old

- **Questions**
  - What’s it like to have EB?
  - What aspects of EB affect you the most?
Self as Different/Normal

The reactions of other people

**Rachel:** I’d just (wish) that people…. wouldn’t be so horrible, like stare at me and things, ‘cos everywhere I go, you just have to get used to it. 
People stare at you

**Rachel:** I wouldn’t want to be treated differently to everyone else ‘cos I am just normal. I just happen to have a skin problem, I am just like a normal person.

**Kerry:** some people might think that I’m not a full person ‘cos I can’t do all the things that other people can do. ‘Cos I’m not like them they think that I don’t count as a proper person.
Being seen as Lazy

- Bella: You’re just sat there and all they can see is someone sitting in a wheelchair... it’s like you’re just lazy

- Kerry: ... they said that I was making it up. ‘Cos sometimes I can walk but some of the time I need a wheelchair ..... And then they started calling me names, like liar and things.”
Self as Different/Normal

Self as Weak

- **Dilip:** I’m weak…(EB) makes me feel different ‘cos other people are strong and I’m not.

- **Adam:** …everybody else can do stuff for longer than me. It’s quite hard. (TEARFUL)

- **Bella:** it feels like you’re ill ‘cos you have to be stuck indoors all day…but you’re not ill, it’s just that your feet are sore…you just want to get out there and do it. But you can’t ‘cos your feet hurt so much.
Self as Different/Normal

The Importance of Belonging

- **Kerry:** (after she attended a DebRA event) I felt happy in a way, ‘cos I knew then that there were other people like me...‘Cos if they have the same thing as me, they might have the same feelings.

- **Caz:** I think there should be kind of a group thing...where there’s other people with, not just EB, but other disabilities...maybe we could go on a trip...something that we could all do together.
Self as Different/Normal

The benefits of having EB

- **Rachel:** …maybe if I didn’t have EB then, I don’t know, maybe I’d be making (sarcastic) little comments, I don’t know…. so maybe it is kind of good that I have it so I know not to do those things. I don’t know, maybe I’d be more different. I could be spiteful or something like that.

- **Kerry:** Some people aren’t very nice to me, (but) I treat them how I would like to be treated, not how they’re treating me.”
Relationships with Friends, Family and Professionals

Self as Burden

- **Matt:** (Mum) would feel more secure without having to worry about my EB all the time, like before sport... sometimes she gets quite worried if I get up in the night... life would be a bit easier if I didn’t (have EB) because then she wouldn’t have to always cope

- **Bella:** ... You have to get other people to do everything and I think they get a bit annoyed... like getting water and (other things) for you. That’s got to be annoying for them.
Relationships with Friends, Family and Professionals

- **Kathryn:** … say my hands or my feet were bad, then we all wouldn’t go (swimming). (My Brother) would be ‘it’s all your fault Kathryn, you can’t do anything!’ Sometimes people don’t say things like that but I think about it, and I think that it’s all my fault…..I think that me getting all the attention kind of pulled me and (My Brother) apart.
Relationships with Friends, Family and Professionals

Relationships with professionals

- **Kerry**: ‘cos I’m nearly 11 now, so I don’t want them telling me everything I can and can’t do when they don’t even know...’cos they’re not me and they don’t have it.

- **Chris**: here (at this hospital) they just fix whatever they have to and they just say goodbye.

Dependence = Defective

- **Rachel**: ...at primary school, teachers could make me feel singled out; they probably thought they were helping me.

- **Kathryn**: At the time I was like ‘yay I’ve got time off school’ but then afterwards I wished I hadn’t done it.
Coping

The importance of succeeding in other areas of life

- **Rachel**: ...when I beat people at stuff, even though I’ve got EB I can still beat them, it makes me feel good....like if I won a test or something like that, if I came highest
Feeling guilty for feeling bad

- **Rachel:** ...I only have EB simplex and I know it’s bad EB simplex.... but still, there’s lots of people who have worse EB and they can’t even eat.... And it just makes me feel a bit guilty.... I do get pain and it’s really painful but I can still do things... I don’t want to patronise them or anything but it’s like that I’m really lucky to not be in that situation.

- **Kathryn:** ...it just makes you feel a bit selfish... half of you is thinking ‘ouch!’ and the other half is thinking, ‘it’s not as bad’.
Conclusions

- In all 3 EB groups, children
  - struggled with the negative reactions of other people
  - struggled to work out how they are different and how they are the same compared to people without EB
  - found that their physical limitations interfered with their ability to join in with their peers/made them feel badly about themselves.
  - felt that they were a burden on their family and friends
Conclusions

- **Children with EB Simplex**
  - were sometimes disbelieved about their special needs
  - felt guilty about feeling bad
Recommendations

- Help children to develop positive self-worth
- Watch out for anxiety and peer/social problems
- Help children to deal with negative reactions
- Pain management
- Psychosocial support for children with all EB sub-types
Overview

These children have proven that regardless of their complex condition, despite increased likelihood of experiencing certain psychological and social problems, that, as a group, they are fairly well-adjusted, and demonstrate several strengths in their ability to cope with a number of physical, psychological and social challenges.

The goal of psychosocial input should be to support and facilitate and to foster individual strengths and resources not to pathologise or to place limits on personal development and aspirations.
If you’d like to discuss these findings more...

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