Quality of Life and Illness Perception in Adult EB Clinic Patients

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- Consultant Dermatologist, Clinical Nurse Specialist, Social Worker, Occupational Therapist, Physiotherapist, Dietician, and Clinical Psychologist

- Role of the Clinical Psychologist:
  - Consultation to EB team in relation to patient management
  - Psychological Assessment
  - Individual Psychotherapy
  - Screening service for QoL and psychosocial stress related to EB
  - Input on research and service development
Rationale for Psychology within Dermatology and EB team.

- There is a complex relationship between skin disease and psychological morbidity.
- Skin diseases affecting patients during childhood/adolescence are proposed to have a significant effect on the formation of trait introversion/extraversion and these traits influence expression of psychological morbidity in relation to skin disease. (Magin 2008)
Rationale for Psychology in Dermatology and EB team

- Typically the psychological morbidity of dermatology patients tends to be minor but impacts on quality of life and may be a risk factor for episodes of depression and anxiety:
  - Self-consciousness
  - Stigma
  - Shame
  - Decrements in self image
Psychological functions of skin

- Touch / tactile experience
- Role of communication
- Mother-infant bonding
- Emotional & physiological regulation of infant and child
- Skin functions as a container to distinguish inside/outside; me/not me
- Physical boundary between self and others/environment is breached
Psychological Impact of EB

- Impact on children – pain, itching, appearance, sense of being different from peers (Van Scheppingen et al., 2008), social isolation (Horn & Tidman, 2002).

- Impact on families – on parents’ relationship as a couple; on parental employment and leisure time; feelings of guilt (Fine et al., 2005; Van Scheppingen et al., 2008);

- Children have been found to be more adversely affected by their EB than adults (Horn & Tidman, 2002; Tabolli et al., 2009), adults have more control over environment and have learned how to cope.
Psychological Impact of EB on Adults

- Considerable impact of EB on quality of life using a postal survey of Dermatology Life Quality Index - similar to moderate to severe psoriasis or atopic eczema. (Horn & Tidman, 2002) (N EBS = 143; DEB = 99; RDEB-HS = 6)

- QoL found to be correlated with the clinical severity of the EB – main issues were being unable to participate in every day activities leading to isolation and psychological problems. (Horn & Tidman, 2002).

- In contrast, a small hospital based survey (n – 20) reported high levels of psychological well-being, normal intellectual, social & emotional development and acceptance of chronic disease (Andreoli et al. 2002).
Psychological Impact of EB

- In a postal survey by Tabolli et al (2009) of EB children and adults in Italy (N=125, EBS =18; JEB =15; DEB = 91, RDEB = 70, KS = 9), patients with higher perceived disease severity had significantly higher scores in all Skindex-29 scales. Psychological components of QoL were no different to normals, and values were low compared to those with other dermatological conditions.

- Gender differences were identified, with females more likely to report a poorer QOL & more likely to have a clinically significant score on the GHQ, indicating possible anxiety and depression.
Psychological Impact of EB

- In 82% of patients, EB had some negative impact on quality of life, although patients tended to adjust well.
- 80% of patients experienced psychiatric symptoms but these were mainly sub-threshold clinically and few required treatment.
- No correlation between severity of EB & level of psychological impact (possibly due to small sample).
Psychosocial Impact of EB

- Qualitative study by Dures (2011) Interviewed 24 adults with EB (EBS, JEB and DEB)
- Key themes that emerged from the analysis:
  - Beliefs about containing the impact of EB affected the experience of EB variably (intrusive to manageable)
  - Understanding the disease well was linked with good management and coping
  - Disabling impact of EB -
    - Physical, Social (stigma)
Outcome of the studies

- Highlighted the psychosocial issues of living with pain, physical impairment, and disfigurement.
- Dynamic nature of the psychosocial impact – the individual experience of illness can change across time and contexts.
- Therefore, support and interventions should focus on how the illness is subjectively experienced.
- Implications for provision of support: informational needs, self-management, peer support, social skills, and individual therapy.
Aim was to document the Irish experience of EB by assessing EB specific QoL so that patient needs could be assessed and addressed.

As previous research showed a significant role for Illness perception on illness experience, this was also assessed as illness views can change over time (Paterson, 2001)

Determine what level of psychological intervention would be acceptable and useful to patients.
Methodology

- Ethical approval obtained from Joint Research Ethics Committee.
- Cross-sectional and longitudinal in design (questionnaires repeated over time)
- Participants – all adults recruited from clinic attendees at the national service for individuals with EB at St. James’s Hospital.
- Participants are already engaged with a service, and so can access psychological input if needed.
Methodology

- Psychology screening at each visit to the specialist EB clinic
- Measures completed:
  - Quality of Life in Epidermolysis Bullosa Q’aire
  - Brief Illness Perception Q’aire
  - Information extracted from healthcare records
- Standard feedback form for EB team
- Individual findings that are clinically significant – a referral made to Psychology
Measures

- **Quality of Life in Epidermolysis Bullosa Questionnaire** (Frew, Martin, Nijsten & Murrell, 2009). Good face validity, acceptable to patients. EB specific.
  - 17 items
  - 4 point answer scale
  - E.g. Does your EB cause you physical pain?
  - How frustrated do you feel because of your EB?
  - Does your EB affect your ability to move about at home?
● **Brief Illness Perception Questionnaire**: (Broadbent, Petrie, Main and Weinman, 2006).

● Illness perceptions are organized cognitive representations or beliefs that patients have about their illness.

● Adjustment to chronic illness involves acceptance of some limitations, and may also require the acquisition of new skills and changes to routine so that the patients can manage the illness. These beliefs have been demonstrated to impact on illness related behaviour, adjustment to illness and outcomes.

   - 9 items each rated on a 0 – 10 scale
   - E.g. How concerned are you about your illness
Preliminary Findings

- Participants were adults attending the EB clinic at St. James’ Hospital (N=36)
- Diagnoses:
  - EB Simplex \( n = 30, \) (83.4%)
  - RDEB & RDEB Inverso \( n = 5 \) (13.9%)
  - DDEB/ EB Pruriginosa \( n=1, \) (2.8%)
- Age: Mean: 33.94, Range 16 - 68
Descriptives – Full sample

- Equal gender distribution Males 17 (47%) Females 19 (53%)
- Mean EBQoL : 11.04 (sd 6.95 range 1 – 32)
- Mean PsychEBQoL 3.85 (sd 2.70 range 0-10.5)
- Mean PhysEBQoL 7.19 (sd 5.11, range 0 – 24)
- Mean BIP: 38.40 (sd 11.80 range 10 – 62)
- Implications: Overall impact of EB on QoL reported as being quite low on average, but some variability.
- Illness perception at mild level generally
Descriptives : DEB

- Equal gender distribution Males 3  Females 3
- Mean score EBQoL: 9.67 (sd 11.36 range 2 – 32)
- Mean score Psych.QoL: 3.67 (sd 2.66, range 1 – 8)
- Mean score PhysQoL: 6.0(sd 9.01 range 1 – 24)
- Mean score BIP 39.83 (sd 14.12 range 25 – 62)
- Implications : overall low level of complaint but high variability in this small group. Illness perception no higher than in Simplex grouping.
Preliminary findings – EB Simplex

- Equal gender distribution
  - Males N = 14 (46%)  Females N = 16 (53%)
  - No gender differences found on total scores of EBQoL
  - No gender difference on BIPQ scores

- Mean score EBQoL : 11.32 ( sd = 5.96 range 1 – 21)
  - Scale range 0 – 51, no published indicators of “caseness” for scale so overall complaint was relatively low.

- Mean score BIP : 38.12 (sd= 11.55 range 10 – 56)
  - Scale range 0 – 80, no published indicators of caseness for scale but higher scores represent more severe illness perception, thus on average illness perception was moderate.
Findings - Gender

- No gender differences found on total scores of EBQoL
- No gender difference on BIPQ scores
Findings: EBQoL

- Psychological/Social items (7) separated from Physical/Practical items (10).
- No significant gender differences, although tendency towards females reporting higher psychosocial EBQoL problems.
- A positive significant relationship was found between Psychosocial EBQoL and total BIPQ score ($r = .56$, $p = .001$).
- No significant association was found between Physical/Practical EBQoL and BIPQ score ($r = .33$, $p = .07$).
Preliminary Findings - AGE

- Age was not significantly associated with EBQoL Total Score, Psychosocial EBQoL, Physical/Practical EBQoL, or BIPQ Total Score.

- This finding held true for the full sample, and the sample with EB Simplex.
Implications of findings

- The findings show that illness perception is not associated with level of physical and practical problems, but is significantly associated with report of social and psychological problems.
- Report of negative illness perception may be a good indicator for brief cognitive intervention with the aim of reducing psychosocial impact of EB.
- Qualitative enquiry showed that the most common problem was feeling frustrated with limitations, and significant impact of EB on social life.
- Age was not a significant correlate of complaint.
- Females tend to report more psychosocial QoL problems than males.
Clinical Considerations

- Patients report what appeared to be a relatively low impact on quality of life, despite the issues involved in living with EB. Perhaps most people are adequately supported and do not require any specific psychological intervention, and have generally adjusted well to this chronic condition.

- As many patients initially denied problems but then revealed more as they became engaged, it seems that patients may use positive denial as a strategy as Margari (2010) already noted.
Clinical Implications

- Experience of setting up a psychotherapy service for EB patients in London – “take up is slow and wary”, despite indications from the team that the need was high (Moss, 2008).
- Uptake of psychology services at our clinic was greatly increased as a result of engagement with screening, so that approximately one third of patients had a brief input during clinic, and four went on to have a longer intervention. Brief cognitive work seems to fit well in the clinic context.
Clinical Implications:

- The questionnaires are brief and have good face validity thus were acceptable to patients.
- The questionnaires are simple to score and may be completed by the patient alone, or with any multi-disciplinary team member. Our experience is that patients divulge more when in an interview context.
Future Directions for research and intervention.

- Need to assess if intervention is associated with improved outcome on BIPQ and enhanced EBQoL.
- Need for longitudinal analyses to measure change over time; anecdotally, it seems that patients’ adaptation may vary over the life span.
- Need to comparisons of QOL scores with similar EB populations in other countries.
- Need to classify severity of disease and compare with levels of QOL.