Life with Epidermolysis Bullosa
Perception and experience of people living with EB and their caregivers

Brief presentation of Results

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Survey carried out for Debra Italia by Doxa MarketingAdvice
Introduction

Debra Italia Onlus
For over 26 years, DEBRA Italia Onlus has supported patients, spread the knowledge and promoted research on Epidermolysis Bullosa in Italy,

Debra Italia is the association who promoted the creation of:
• two main multidisciplinary EB centres in Italy (in Rome and Milan, now both in the ERN network)
• monodisciplinary excellence centres in Catania (hand surgery) and Turin (dental care)

In the last year particularly, Debra Italia Onlus has acheived some important objectives:
• promoted the inclusion of Italian EB patients in International trials,
• increased international presence in Debra International and in other international rare diseases organizations
• renewed the image of the association with marketing activities, with a new logo, new social networks, new Internet site, the first advertising Video ever realised in Italy and new marketing collaterals
• Increased Debra Italy and EB visibility on media channel
• massively increased the number of national associate members and assembly attendance

In parallel with these activities DEBRA Italia has commissioned a survey to give voice to EB patients and their most direct caregivers by undertaking an in-depth path of research, knowledge and understanding of their perspective
Survey objectives

Survey has been carried out by Doxa and their specialised professionals

Objective:
• collect people living with EB direct and spontaneous experience not only with the disease but also and most of all with the care and daily management of the symptoms

• obtain precious indications to understand the patients’ experience and perception,

• further develop the specific guidelines for handling this insidious disease;

• to give greater social resonance, media visibility and tangibility to the world surrounding EB.
Research design

The survey tool was

- **in-depth interviews** conducted with a **non directive qualitative approach**, aimed at collecting the respondents' **spontaneous experience**.

- **telephone calls**: considering patients are spread-out on the whole Italian territory, the telephone channel was adopted. The interviews lasted 60 minutes on average.

The people involved in the study:

- **33 people** all over Italy who made themselves available after a direct invitation by Debra Italia

- **overall we heard the story of 18 adult patients and 15 caregivers**, allowing us to explore **30 EB cases**.
From the analysed sample emerged that:

**SIMPLEX**: does not have an excessive impact on life's quality, allowing patients to lead a completely autonomous life.

**JUNCTIONAL**: a quite homogeneous group in terms of symptoms → all patients can use their hands and have a good level of overall autonomy.

**RECESSIVE DYSTROPHIC**: has the widest range of clinical manifestations and the greatest heterogeneousness in terms of being overall compromised. Most patients suffer from the progressive fusion of the fingers, an aspect which is acknowledged by all as extremely incapacitating.

**DOMINANT DYSTROPHIC**: the symptoms are quite moderate, patients can expect a normal life without any impact on a regular physical development.
Survey subjects

- **Experience with the disease** ➔ the impact of EB on the patients’ life and the main critical areas, defined and described with the words of the people who live with it.

- **The different areas of everyday management of the physical impact of the disease** ➔ wound treatment, pain, hygiene, nutrition, clothing.

- **The concept of independence and self-fulfilment** ➔ in the private and domestic life, at school and work, during leisure or cultural activities.

- **The patients’ self image** ➔ how they perceive themselves and their sense of efficacy in the world, what are their recurrent thoughts.

- **The patients’ social image** ➔ distinguishing between the family dimension and the larger social one (school and friends).

- **The social dimension** ➔ seen by parents both in the family perspective and in a broader one, in terms of development of their child’s social relationships.

- **Perception of the future** ➔ how patients see themselves in the future and how their caregivers see them: what is the desire and the expectations.
The survey generated a huge amount of information:

- **100 CA. PAGES REPORT**: the interviews generated a report including almost 100 pages of information, and spontaneous quotes from the sample.

- **QUALITATIVE RESEARCH**: qualitative data, a few numbers.

- **NEXT STEP**: Quantitative phase on 100 people.

This brief presentation includes only a few information taken out from the overall results.
Experience with EB
Overall considerations

These are the very first dimensions caregivers and patients spontaneously associate with the disease:

- **FRAGILITY** → the first recognizable characteristic in descriptive terms is fragility, of the skin but also of the patient as individual in general.

- **UNPREDICTABILITY** → and the associated impossibility to plan, both in the short and long term, due to the high incidence of accidents and complications. The condition is one of constant alert in both the everyday routine and in handling unexpected events.

- **DEPENDENCE** → the most incapacitating aspect and one of the most difficult to accept for patients, the origin of the weight of responsibility for caregivers. Mutual patient-caregiver dependence

- **PAIN** → the patients’ suffering and the caregivers’ sorrow due to their sense of helplessness
Pain: a profoundly different perception

A central and truly disrupting aspect emerged from the survey lies in the profound difference characterizing the role attributed to pain by patients and caregivers. To understand its meaning, three levels must be distinguished:

1. **THE MEDICAL-SCIENTIFIC LEVEL (control)** → which confers a key and peculiar role to the control of pain and its peaks in this disease.

2. **THE FAMILY LEVEL (helplessness)** → Pain represents the first concern for caregivers who helplessly witness the multiplication of the wounds on the child's body and they associate them with a pervasive experience of pain. They are supported by the clinical literature and practice on this topic but they are unable to fully understand the perceptive entity of this pain.

3. **THE PERSONAL LEVEL (tolerance)** → namely the patients' authentic and real experience with pain. Patients are born, grow and live immersed in this condition, thus developing a tolerance scale that is extremely different from the people who take care of them.

For this reason, because it's a congenital physical condition for them and they have always lived with it, patients mention pain less frequently than caregivers, as if it was a secondary limitation compared to the structural restraints imposed by EB, which represent the true limit.
Autonomy

For patients, the structural restraints imposed by EB represent the true limit.

EB patients with a dystrophic or junctional type are never fully autonomous in handling their disease.

FOR WOUND TREATMENT → they always need help, if not continuously at least in the preparation of medications.

FOR OTHER DAILY ACTIVITIES → the patients' autonomy largely depends on the extent to which their hands’ functionality is compromised.

- **DEPENDENCE** → the most disabling aspect and identified as one of the most difficult to accept for patients.

- **FROM AUTHONOMY TO EMANCIPATION** → the importance and desire of acquiring not only physical independence but also and most of all a psychological emancipation (benefits of work: those who have a job said it has beneficial effects also on their health).
Experience map

1. To accept
   EB is part of me and I face it

2. To endure
   EB is stronger than me, it dominates me

3. To celebrate
   EB is a gift, it makes me better

4. To deny
   EB is not a problem, it has no impact on me

The rational approach results in two ideal-typical profiles: 1. some accept EB, show great skills in handling it and are inclined to face the limits it imposes in a constructive way 2. others underline the constant difficulty in handling it and the heaviness of this condition (this area includes a good number of caregivers).

The emotional approach results in the following ideal-typical profiles: 3. in order to learn to live with it, there are people who describe EB as a privilege, a condition that allows them to have access to higher spiritual and emotional values. 4. on the other hand, there are those who tend to deny the difficulties it creates and neglect EB, with more or less serious effects on their quality of life.
The distribution of patients on the map

1. To accept
   EB is part of me and I face it

2. To endure
   EB is stronger than me, it dominates me

3. To celebrate
   EB is a gift, It makes me better

4. To deny
   EB is not a problem, it has no impact on me
The distribution of caregivers on the map

1. To accept
EB is part of me and I face it

2. To endure
EB is stronger than me, it dominates me

3. To celebrate
EB is a gift, it makes me better

4. To deny
EB is not a problem, it has no impact on me

Caregivers
The future
Respondents wish list for the future (with their words)

All expectations mentioned can be grouped in six areas:

1. All respondents wish for a widespread increase in the level of knowledge of the disease, both on a technical, medical and on a social level (awareness);

2. Our respondents also expect improvements/steps forward of scientific research on very practical clinical aspects
   - The possibility to prevent the formation of blisters
   - The possibility to avoid the detachment of the skin on the blisters
   - The management/prevention of hands' malformations and a greater efficacy in reconstructive surgery
   - A greater wearability and comfort of fingers' braces (splint)

3. The expectations for improvements on specific aspects of NHS (beaurocracy)

4. Expectations on clinical centres (more centres and closer to them)

5. Architectonical barriers

6. Expectations from Debra: what should the association do to better support the life of people living with EB (vehicle information to families and advocacy with authorities)
Next steps
Next steps

How the survey is being used:

**Address scientific – psychosocial experts community**

- Sharing of results for scientific/psychosocial specialists supporting EB patients (whitepaper or publication?)

**To increase awareness of EB on general public**

- Press conference on 14th September in Milan (with great response from the press)
- Press release
- Social network

**To improve Debra’s activities**

- Source of insights for next Debra’s actions to support EB people
- Precious source of communications targets and language
- Follow up with a quantitative research