THE IMPACT OF LIVING WITH EPIDERMOLYSIS BULLOSA:
SUMMARY OF THE INTERVIEW FINDINGS

The reason for the study:
This study is one of a series of DebRA funded research projects exploring the psychosocial impact of living with EB. The series of studies is being done because there is evidence from both people with EB, and the healthcare professionals who work in the field, that some people affected by EB can face a combination of practical, social, and psychological difficulties in the course of their daily lives, which result in needs that are not being met. Research is needed to clarify the extent and type of these needs, so that effective provision strategies can be developed. Research is also needed to look at the ways in which people feel they manage their EB well, and the available support provision that they find helpful. This is in order to learn more about successful management as well as the difficulties, and to try and ensure that current good practice continues.

There were 2 main aims:

1. To understand more about the ways in which EB can affect everyday life and how people manage the condition.

2. To gather information on existing support that people use and value, and support needs that are not currently being met.
Overview of the findings:
The findings in this report are based on a series of one to one interviews with adults who have EB. The interviews were conducted face to face, over the telephone, or on-line. In total 24 people (10 men and 14 women) with the 3 main EB types (EBS, DEB, and JEB) took part.

The findings are based on my interpretation of participants’ words, after speaking to many people who expressed a range of thoughts, feelings, and experiences. Although everyone who took part in the interviews gave a unique account of living with EB based on their own experiences, this report looks at some of the common issues that participants talked about. Although in reality these issues are difficult to separate from each other, for the sake of clarity I have grouped them under broad headings:

1. Personal beliefs about the impact of EB in everyday life
2. The effects of EB on the family
3. Experiences of managing EB
4. The disabling impact of EB
Summary of the ways in which EB can affect everyday life:

Personal beliefs about the impact of EB in everyday life:
Views range from those participants who find that EB intrudes into all areas of their lives through to those who feel that they are able to work around the condition and limit its impact. The person’s sense of control and the manageability of their EB in everyday life influence how much the condition is affecting their quality of life.

Personal beliefs about the extent to which the effects of EB can be controlled and limited can change over time. From day to day, season to season, and year to year, managing EB can become a more or a less central part of daily life. Participants spoke of a variety of responses to living with EB. Responses which participants feel are positive and work well for them include taking each day as it comes, a belief in their ability to cope and find a way round things, and a sense of feeling lucky because things could be worse. Responses which participants feel have a negative effect include feelings of anger and frustration, bottling up negative emotions, a loss of self confidence, and the urge to withdraw from other people.

The ability to recover from setbacks was seen as important by participants in relation to their belief that they can continue to meet the challenges of EB. There are variations in how well participants feel they have been able to adapt to, and overcome, the difficulties of living with EB. Some people with EB feel that there are positive consequences of the challenges that they cope with in their lives. However, others cannot always find the strength to stay positive. People find that their ability to bounce back from difficult times can alter. For example, some participants described periods when they felt less able to deal with their EB, but they then went on to recover a greater sense of control and acceptance. Things that can affect the impact of EB at particular times include changes in symptoms, personal circumstances (for example relationships) and developmental stages of life (for example being a teenager).
The impact of EB on the family:
Many participants spoke of how well their family members adapted to the birth of a child with EB, and of the great source of support that families continue to provide. However it was also acknowledged that in some families parents found the pressures harder to adjust to. Some participants feel that this has affected their relationship with their parents. In some cases nursing and skin care routines seemed to take over daily life. In discussions of bringing up a child with EB, many adults with the condition feel that children should not be ‘wrapped in cotton wool’, and believe that being allowed to learn from experience is a positive approach with long term physical and psychological benefits.

In families where children have EB, parents will usually provide nursing care. For some people with the condition having a parent as a carer, works well and provides a good source of practical and emotional support through childhood and into adulthood. But even in families where parental involvement is very positive, some participants did not want these caring roles to continue into adulthood. They expressed concern that EB can dominate family life if caring responsibilities stay entirely within the family.

Some participants expressed a sense of shared experiences and empathy with other family members who also have the condition. By contrast some participants find that family members who do not have EB cannot fully appreciate what it is like to live with the condition.

The effects of EB on children in the family were talked about. Some participants with the recessive forms of EB explained that their children have experienced hurtful teasing because of their parents’ condition and visible difference. For some adults with the dominant forms of the condition, their first hand experience of the effects of EB is a source of concern about what the future holds for their children.
Experiences of managing EB:
Participants talked about living with a lifelong and non-curable condition, and how it can be hard to think about the future and the ways that EB may affect them. In some cases EB can be life-shortening. Whilst this is an issue that people did not talk about very much, an awareness of the possibility of developing EB related complications, such as squamous cell carcinomas (SCC), was expressed. Concerns about the longer term impact of EB seem to increase with age.

The genetics of EB raises issues for many individuals. One concern is having children, and the possibility that they may inherit the condition. Some of those interviewed feel that information about the options available to people with EB is not always clear. Some participants with a recessive form of EB also talked of the guilt and shock that their parents experienced on learning that they had unknowingly passed on the condition.

Another important aspect of EB that was highlighted is that the condition can be unpredictable and variable. Despite having the condition all their lives participants often cannot know how their skin will be at any given moment. Some adults with EB have lived with the added uncertainty of not having a clear diagnosis, which has meant coping with an unrecognised condition. This can lead to practical challenges, such as not being in a position to make informed decisions that may help them to manage their symptoms. There is also the psychological impact of not knowing what was happening to their skin or why, and of feeling different from other people.

People with EB have to consider how activities will affect their skin. Choosing between the pain of doing an activity versus the frustrations of not doing it becomes part of everyday life. An added complication that participants spoke of is that when they find successful strategies for managing their EB, other people sometimes find it even harder to appreciate the challenges that they face.
Several participants explained that an important aspect of self management is understanding that everyone’s skin is different and that each person has to know their own body. This can be the case even within families, where members who share the same condition need to recognise that each individual may be differently affected. Understanding this helps people to have confidence that they know what works best for them.

Living with EB often means meeting a lot of healthcare professionals. Many participants have very positive experiences of the specialist services provided by DebRA. By contrast, there were many examples given of GPs and other healthcare professionals who did not know about EB, or who were negative and unhelpful. There have been times when participants have felt powerless in interactions with medical people and believe that they experienced unnecessary physical and psychological suffering due to a lack of understanding. Life long experience also means that sometimes people with EB feel more knowledgeable than some of the healthcare professionals they meet, which can cause difficulties. The ability to talk to professionals assertively and clearly is seen by many as important for receiving quality healthcare. Consequently participants value good working relationships with healthcare professionals, and continuity of care.
The disabling impact of EB

The symptoms of EB, such as pain and fragile skin, can be physically limiting. As a consequence participants can sometimes feel left out from ‘normal’ everyday activities. This can be made worse by society’s attitudes to physical impairment, visible difference, and skin conditions.

Other peoples’ lack of understanding about EB can make life more difficult. Sometimes when participants try to explain about the condition and the restrictions it causes, people don’t believe them or dismiss what they say. This is particularly the case when the effects of EB are not visible.

When people with EB are out in public, they sometimes have to cope with negative reactions to their skin and appearance. These include staring, unpleasant comments, and discrimination (e.g. not being allowed in public swimming pools). Some participants have developed ways of coping with hurtful attitudes, but others find such experiences “soul destroying”.

Some people with EB feel dissatisfied with their appearance due to the limitations of what they can comfortably wear. People explained how not being able to dress the way they want can increase their feelings of difference and isolation from other people.

Many participants conveyed how EB affected their education. Sometimes it was due to symptoms, such as pain and time off due to blistering. Other times it was due to the school environment, for example bullying and a lack of understanding about EB. These experiences have often affected participants socially and emotionally, as well as educationally.
Employment can present a range of challenges. Some participants have chosen to be registered as disabled, although this process is not always straightforward. Generally, people with EB working in supportive environments have found that informing colleagues about their condition has enabled them to adopt routines that help them to manage. Several participants reported positive psychological and social effects of being able to work. By contrast, other participants experienced negative and discriminatory reactions in the workplace. Some participants expressed worry about being stigmatised because of their skin condition, and therefore choose not to reveal their EB. Those participants who are not able to work often face the challenge of going through the benefits system. The difficulty of explaining EB, particularly if someone is unsympathetic, can be distressing and frustrating.

Finally, living with EB has a financial impact on participants and their families, for example by having to pay for your prescriptions. This is an aspect of the condition that many of those I spoke to find unfair and unacceptable.
Summary of existing support and unmet support needs:
The following summary is based on participants’ views about the current support which works well, and the areas where support is lacking. Many of those I spoke to stressed that there cannot be a ‘one size fits all’ approach to providing support, and that needs will vary from person to person, and change over time.

Sources of support:  For many people with EB, family and friends provide a great amount of practical, social, and emotional support. The specialist nursing and social care team services offered by DebRA are another important source of help, as are local healthcare and social services. Participants also draw on their own inner strengths and knowledge for support. It is difficult to separate out what these different sources of support provide and how they affect peoples’ quality of life. However, I have grouped participants’ views on support into 3 broad areas:

1. Support from professionals
2. Peer support
3. Strengthening people’s inner resources
Support from professionals

DebRA services: Many participants expressed the view that DebRA nursing and social care teams provide excellent and effective support. DebRA specialists are valued for their expertise and their caring, dedicated approach. However not everyone I talked to accesses these services. One reason is the widely held belief that DebRA’s main focus should be on caring for people who have the most clinically severe EB. As a result some of people, especially those with EBS, feel that they should not use DebRA resources. Whilst some of these people are able to manage well without help, others experience practical, emotional, and social difficulties, and would like to have more direct involvement with DebRA services.

Information needs: Participants talked of the need for easily and freely available information on a range of topics. For example, some participants feel there is a lack of information for people who do not have EB themselves but are close to someone who does, such as friends and partners. Such information could cover issues such as becoming intimate and having sexual relationships. Any material should explain what it can mean to live with EB and should be presented in an accessible, informal style, rather than in detached medical terms. This would allow people who do not have EB to gain information without having to rely on the person with EB to explain everything.

Many participants would like information about the genetic implications of EB, for example a clear diagnosis of their EB type/subtype, and the likelihood of passing on the condition to future generations. People would also welcome information on accessing genetic counselling services and on the tests that they currently offer.

Related to this is a desire for up to date information on clinical research. Whilst participants generally support DebRA’s financial investment into searching for treatments and cures, there is some frustration they are not told about the progress that is being made, or how and when medical advances could affect their daily lives.
**Support for family:** Participants talked about the great impact of EB on the family. Whilst some people feel that their family have always been well supported, others feel they have had to cope with very little help. People expressed the view that practical resources and advice for parents could help restore balance within the family and shift the focus away from EB. One example is how outside help with skin care could reduce the negative impact that carrying out painful nursing can have on the parent/child relationship. Some participants felt that parents may benefit from emotional support, not only to help them cope with their own anxieties about their child’s health, but also with the reactions of others around them to EB. As adults with EB, participants also recognised that their children may have support needs, for example if they become fearful for their parent’s health. However many participants also feel that there is probably more support available nowadays, than when they were growing up.

**Local support provision:** Satisfaction with local support provision is mixed. In terms of education, healthcare and benefits, experiences are negatively affected by a lack of knowledge about EB. It is recognised that EB is a rare condition so most people are unlikely to understand it. At present the DebRA team often help individuals and families inform professionals, which is a service that many participants value. However it was also felt that information that is specifically aimed at the range of professionals that many people with EB and their families will encounter should be more readily available.

**Peer support**

**Peer support:** Many participants find the chance to talk to other people with EB helps alleviate the sense of isolation that can come from having a rare condition. Peer support is often organised through DebRA, for example with regional meetings or the recently formed MAD (Making a Difference) group. In some cases this is already a valuable service, and one that should be built on. However, other participants feel that they are not always informed about what is happening, and what they could be involved in.
**On-line support groups:** Services are not equally available, accessible, or appropriate for the range of people who live with EB. In particular, younger participants expressed interest in being able to talk to each other on-line, including real time chat rooms.

**A mentoring or buddying system:** Some participants feel that a mentoring or buddying system could provide a good source of support. The idea would be that adults who have lived through transition periods, such as adolescence or starting work, share their experiences and expertise with younger people and their families.

**Strengthening people’s inner resources**

**One to one support:** The ways in which people manage living with EB vary. Whilst some of those I spoke to do not feel the need for outside support, other participants would like some help at certain times in their lives. One example would be the opportunity to talk openly to someone impartial, such as a counsellor. Some participants have found that the help is not available when it would be most beneficial because services are difficult and slow to access.

**Self management and social skills:** Some participants feel that one of the roles of the nursing team should be to encourage people to become the experts in knowing their bodies and managing their condition themselves. This could help give people a sense of control over their lives. It was suggested that this could be done from an early age, for example by organising workshops for children with EB that focus on management skills, as well as providing a chance to meet others who face similar issues. Related to this is the need that some participants expressed for help in dealing with other people. This includes learning how to talk confidently to professionals, such as healthcare workers, so that the person with EB feels less vulnerable. It also includes ways of coping with negative reactions from others, such as staring or unpleasant comments from the general public. Some participants feel that having ideas and input about how to respond in social situations could help improve their self confidence.