



# Living with Butterfly Skin

Epidermolysis Bullosa (EB)



# About Butterfly Skin

## What is Butterfly Skin?

The medical term for Butterfly Skin is Epidermolysis Bullosa, or EB for short.

EB is a group of incredibly painful genetic skin conditions that cause the skin to blister and tear at the slightest touch. People living with EB have skin as fragile as a butterfly's wing, hence the name Butterfly Skin.

In the most severe cases EB can be very visible and can affect multiple areas of the body, however in other cases such as with EB Simplex, (70% of all cases), it can be less visible and affect only certain areas of the body such as the feet. In every case though, EB leads to excruciating pain and can have a devastating impact on the patients physical and mental wellbeing.

EB is not contagious.

## What causes EB?

People with EB have a faulty gene which is passed down through families. The faulty gene means the skin cannot bind together, so any trauma or friction can cause painful blisters, open wounds, and scarring.



## How many types of EB are there?

There are 4 main types:



### EB Simplex (EBS)

The most common type of EB, which can range from mild, with a low risk of serious complications, to severe. With EBS the missing protein and fragility occurs within the upper layer of the skin (the epidermis).



### Dystrophic EB (DEB)

DEB can range from mild to severe (recessive or dominant) with the missing protein and fragility occurring below the basement membrane within the superficial dermis. 25% of all EB cases are DEB.



### Junctional EB (JEB)

JEB is a rare, moderate to severe form of EB where the missing protein and fragility occurs within the structure that keeps the epidermis and dermis layers together - the basement membrane. 5% of all EB cases are JEB.



### Kindler EB (KEB)

Named due to the defective gene being responsible for the information required to produce the protein Kindlin1. This type of EB is very rare but fragility can occur at multiple levels of the skin.

## Which part(s) of the body can EB affect?

In the less severe types of EB, it affects the feet only, however in the most severe cases it can affect any part of the body including blistering on the eyes and on internal organs including the throat. In every form, EB causes lifelong pain.

## How many people does EB affect?

The exact number is unknown as often the condition goes undiagnosed, but it is estimated that there are at least 5,000 people living with EB in the UK, and 500,000 worldwide.

## Are there any treatments or cure(s) for EB?

There are currently no cures for EB. However, treatments are available which can help ease and control the symptoms such as pain and itch. They can also help avoid skin damage, reduce the risk of developing complications, such as infection and malnutrition, and improve quality of life.

## Is EB fatal?

Some of the less severe forms of EB can improve with age, however the most severe forms have a high death rate.



Genetic condition



Skin as fragile as a butterfly wing



Layers of skin tear and blister



Can affect internal organs



Affects an estimated 500,000 people worldwide



Not contagious



No cure



## Living with EB

Isla, 14 from the Scottish Highlands, is living with Recessive Dystrophic EB (RDEB)

“

I get up, I go to the toilet and even that is a struggle, I can't dress myself. I am at my worst when I am having my bandages changed, the pain is unbearable, it makes me scream and I must take strong painkillers every day. I get blisters on the inside, on my oesophagus where a balloon is inserted down my throat and blown up whilst I am under general anaesthetic to get my throat back to its normal width. I still have fingers, but the skin has formed over them. I do wonder what I would be doing if I didn't have EB but there is no point wondering when this is what life has dealt me. I do sometimes think “**why me**” though.”

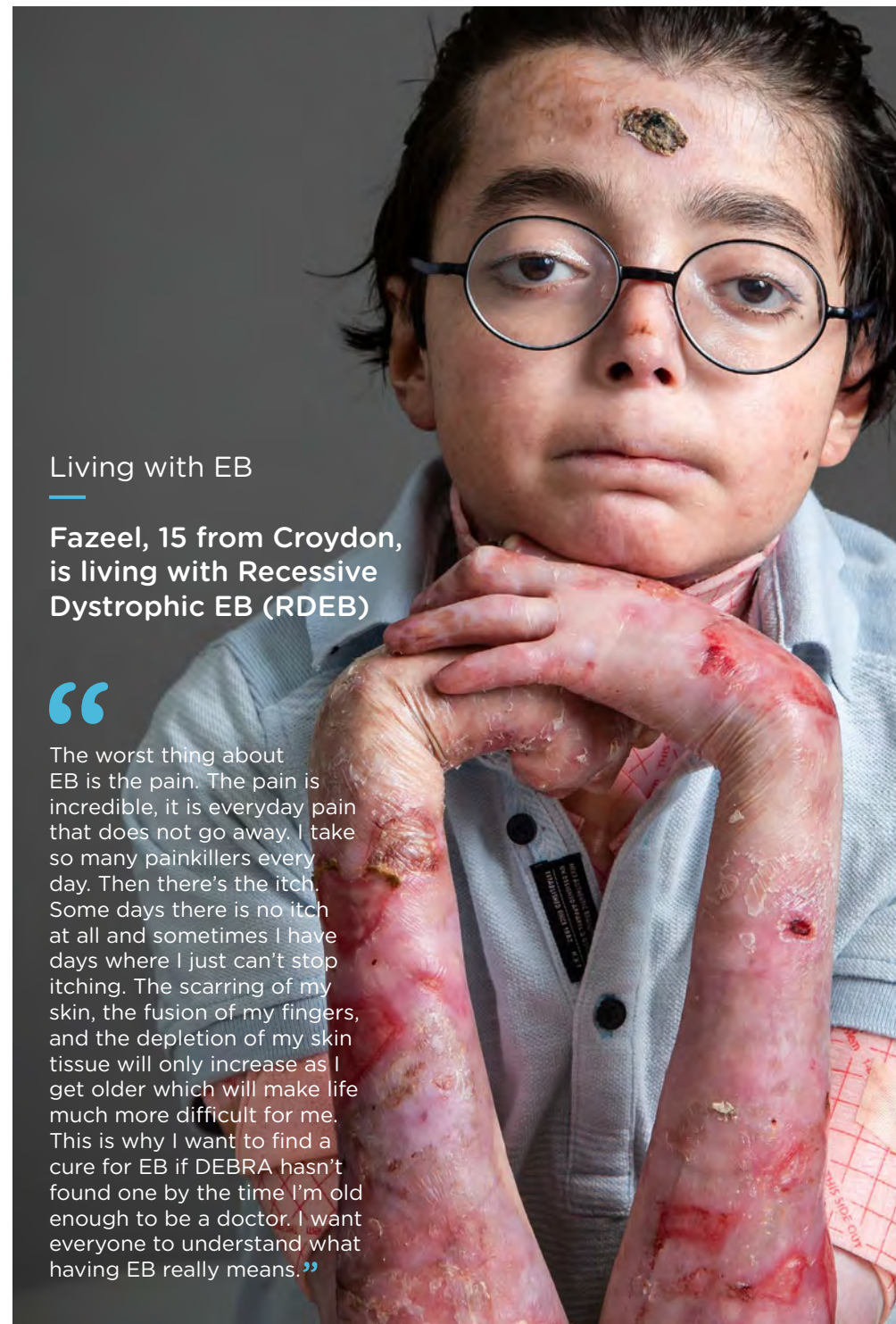


## Living with EB

Fazeel, 15 from Croydon, is living with Recessive Dystrophic EB (RDEB)

“

The worst thing about EB is the pain. The pain is incredible, it is everyday pain that does not go away. I take so many painkillers every day. Then there's the itch. Some days there is no itch at all and sometimes I have days where I just can't stop itching. The scarring of my skin, the fusion of my fingers, and the depletion of my skin tissue will only increase as I get older which will make life much more difficult for me. This is why I want to find a cure for EB if DEBRA hasn't found one by the time I'm old enough to be a doctor. I want everyone to understand what having EB really means.”





# What we do

**DEBRA is the national charity and patient support group for people living with or directly affected by EB.**

DEBRA does not receive statutory funding and relies on the income generated by its charity shops and fundraising activities to provide care and support for people living with EB, and to fund pioneering research to find effective treatments and ultimately, cure(s) for EB.

**With your support we can fund:**



## **Specialist healthcare**

DEBRA works in partnership with the NHS to deliver an enhanced EB healthcare service which is vital for people living with EB. There are four designated EB centres of excellence around the UK providing expert specialist EB healthcare and support, as well as other hospital locations and regular clinics which aim to provide EB services to people wherever they are located.



## **Member services**

The DEBRA Community Support team works with the EB community, healthcare, and other professionals to improve quality of life for people living with EB. They offer support, advocacy, information, and practical help at every stage of life. The DEBRA Membership scheme includes holiday home respite, grants, and bespoke events to support people living with EB.



## **Pioneering research to find effective treatments**

DEBRA supports research programmes that aim to find treatments that will slow, stop, or reverse the progression of EB. Repurposing drugs which are already available and successful in treating other inflammatory skin conditions is a key part of our research programme.



## **International collaboration**

International collaboration and coordination are vital for sharing expertise and developing effective treatments and potential cures for EB. It allows us to coordinate research grants, fund international training, and share expertise and best practice.

## A message from DEBRA Vice President, Graeme Souness

“

From the time I have spent with Isla and her family, I have seen first-hand the extreme pain this devastating condition causes and the daily challenges it creates.

Please join me in supporting DEBRA so that we can find effective treatments to help stop the pain of EB for Isla, Fazeel, and thousands of other people who currently must live with this awful condition.

Thank you.”





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## Corporate partnerships



Get in touch to see how partnering with DEBRA can be mutually beneficial.

# Ways to get involved



## Donate online

To help DEBRA stop the pain of EB, please visit:  
[give.as/DEBRASStopThePain](http://give.as/DEBRASStopThePain)



## Leave a legacy

Leaving a gift in your will is a simple, but vital way to #FightEB



## Join our weekly lottery

Your chance to win up to £25,000 alongside helping #FightEB



## Spread the word

Help raise awareness of EB and DEBRA by simply talking about it



## Join an event

Check out our upcoming events and challenges



## Visit us instore

Donate, shop or volunteer and help make a difference



## Give us a follow

Keep up to date with what we've got going on and follow us on social media