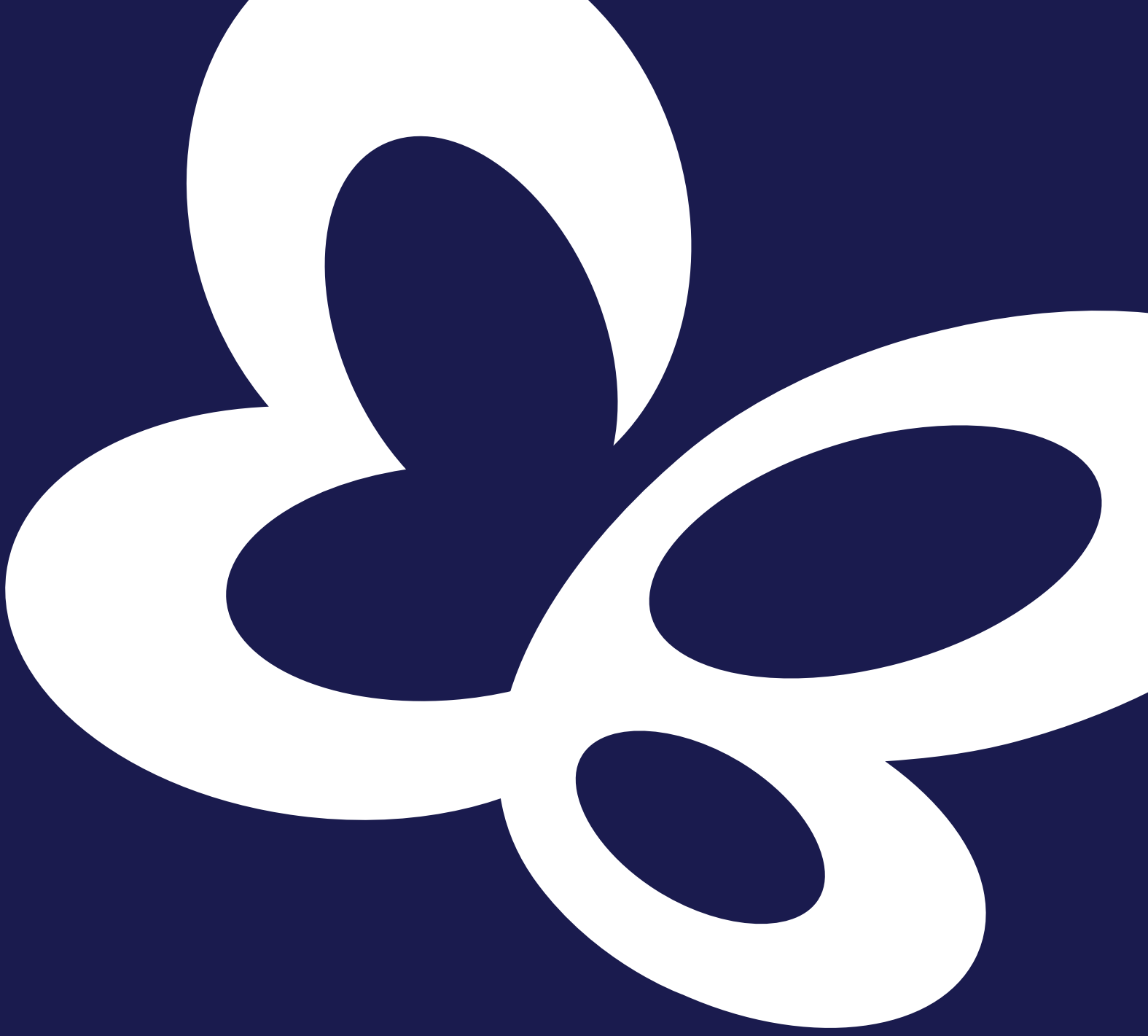

Epidermolysis Bullosa

A Handbook for EB Patients and Families

Developed by the Section of Dermatology at the Hospital for Sick Children with the support of DEBRA Canada

Handbook Development Team

Editors

Elena Pope, MD, MSc, FRCP(C)
Associate Professor of Pediatrics, University of Toronto, Toronto, Canada
Head, Section of Dermatology, Division of Paediatric Medicine, Hospital for Sick Children, Toronto, Canada

Irene Lara-Corrales, MD, MSc
Associate Professor of Pediatrics, University of Toronto, Toronto, Canada
Pediatric Dermatologist, Hospital for Sick Children, Toronto, Canada

Maya Deeb, HBSc
Medical Student, University of Toronto, Canada

Authors

Carmen Liy-Wong, MD
Asim Ali, MD, FRCS(C)
Nimrita Aujla, MSW, RSW
Lauren Briggs, BSc
Fiona Campbell, BSc, MD
Ali El Hamlouly, BMSc
Peter Judd, BSc, DDS, D Paed, MSc, FRCD(C)
Peter Kannu, MD, PhD
Melanie Kirby, MD
Irene Lara-Corrales, MD, MSc
Jamil Lati, MScPT, BScPT, HonBSc
Lisa Lazzarotto, BSc(OT), BA
Michelle Lee, RN, BScN
Margaret Marcon, MD, FRCP(C)
Inez Martincevic MSc, RD
Elena Pope, MD, MSc, FRCP(C)
Hamza Sami, HBMSc
Andrea Shugar, MS, CGC
Jennifer Tyrrell, RN, MN

Graphic Design & Illustration

Maya Deeb, HBSc

SickKids®

debra
Working for a life free of pain.
Pour une vie sans douleur.

DEBRA Canada

Dear Family,

The DEBRA Canada Board of Directors would like to let you know that you have our full support. Our Board of Directors are made up of many patient member families like yourself who understand the challenges you are facing. Please feel free to reach out to us anytime by phone: 1-800-313-3012 or via email at: debra@debracanada.org.

What is DEBRA?

DEBRA Canada is a voluntary, non-profit registered charitable organization dedicated to providing support for people affected by EB and their immediate families; and to increasing Canadians' awareness of this challenging disease.

What does DEBRA Canada do?

DEBRA is the only organization body in Canada exclusively committed to the care and support of families and to improving their quality of life.

Become a DEBRA Canada Member!

In order to join the DEBRA Canada patient member family and access the programs and services provided; please sign up for membership by filling out the DEBRA Canada Membership form included in this book, or online: http://www.debracanada.org. Please note, as per the membership form, you must be a permanent resident of Canada to apply for membership.

Our Programs and Services for Member Families include?

EB Fellowship Program / EB Advice – An initiative of DEBRA Canada and the Dermatology department at The Hospital for Sick Children (SickKids), this program aims to help support patients and families affected by EB throughout Canada. EB.advice@sickkids.ca is an email available to both health care providers as well as EB patients and their families. This program helps bridge the gap in knowledge and patient care and assists in bringing together medical doctors across Ontario and the rest of Canada with the goal of increasing best practices and a higher quality of care for EB patients. For more information on the program, visit: http://www.debracanada.org

Free Infant Care/ Patient Care Kit Program – One of the ways DEBRA Canada supports our patient member families is through the infant care/patient care kit program. Our kit provides a comforting support package for newborns and their families as they begin to care for a child with EB. The kit can be tailored to older patients, not only infants. If you have not received a kit from your EB clinic, we welcome you to contact the DEBRA Canada office to request your free kit.

Medical Assistance/Financial Assistance Fund Program – DEBRA provides financial assistance to families for expenses related to the care and treatment of someone with EB. The request must serve

a medical and/or quality of life purpose. Please see the form included in this book, or go online to download here: http://www.debracanada.org.

Conference Funding for Patients & the Medical Community – DEBRA provides funding to EB patients, health and medical professionals to attend national and international conferences to further increase their knowledge toward best practices and EB patient care in Canada.

Fund a Butterfly Ambassador Wish Program – The DEBRA Canada 'Fund a Butterfly' Program involves the granting of a personal wish to an EB patient (adult or child). The successful applicant will then work as an Ambassador for EB and DEBRA Canada to further the quality of their life, as well as inspire and empower other EB patients to pursue their dreams/goals in the face of adversity. Visit our website to download the PDF application form: http://www.debracanada.org.

Support for Three Canadian EB Clinics – DEBRA Canada provides support and funding to the three EB Clinics across Canada. The EB Clinics are located at: The Hospital for Sick Children in Toronto, ON. CHU Saint-Justine Hospital in Montreal, QC, and the BC Children's Hospital in Vancouver, BC.

Ambassador Program – Our Ambassadors are DEBRA Canada patient members who inform and educate the public about EB, and create awareness about EB year- round. Contact the DEBRA Canada office if you wish to become a DEBRA Canada patient ambassador.

Please visit our website at http://www.debracanada.org for more information.

We look forward to assisting your family.

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Introduction

Understanding and managing a complex disease like Epidermolysis Bullosa (EB) is a challenge for patients and their families. In this time and day families can be overwhelmed and mislead when seeking information. Social media, the internet, Dr. Google, among others provide vast amounts of material that are many times difficult to understand and that don't apply to different types of EB. Finding reliable information in the internet that helps and guides in the care of EB patients is an enormous task and it is sadly not uncommon families are misguided by online sources of information.

Our team in the EB clinic at the Hospital for Sick Children in Toronto and Debra Canada have identified the need to have accessible, reliable and easy to understand information for EB patients and their families of common problems EB patients encounter. We came up with the idea to approach experts in different aspects of EB to create patient and family friendly handouts that explain common problems and their management in EB patients. We want this information to be easily accessible in the hospital's as well as in Debra Canada's website and we also want to have printed copies for EB families. We hope this initiative fills the identified gap in access to information.

Irene Lara-Corrales & Elena Pope
Handbook Editors-in-Chief

EB Clinic at Sickkids

The SickKids dermatology clinic created the first Epidermolysis Bullosa (EB) Clinic in Canada in 2004. Currently, the clinic population followed at SickKids has grown and there are two half-day EB clinics per month and consultations are provided to patients all over Canada.

Clinic Day Structure

Morning	Coordinated appointments in various departments within the hospital if feasible
Lunch	Held in the Dermatology Clinic (Clinic 7), provided by DEBRA Canada
Afternoon	Multidisciplinary EB Clinic

Medical Directors

Dermatology	Dr. Elena Pope
Dermatology	Dr. Irene Lara-Corrales

Main EB Team

Dermatology, Nurse, Clinic Coordinator	Michelle Lee
Dermatology, Nurse	Jackie Su
Dermatology, Social Worker	Nimrita Aujla
Information Coordinator	Kit Lee
Clinic Clerk	Ann Roussos
Dermatology Training	Fellows and Residents

Subspecialties (as needed)

Bone (Endocrinology)
Cardiology
Dentistry
Genetics
GI/ Hepatology & Nutrition
Haematology
Image Guided Therapy
Nephrology
Occupational Therapy
Oncology
Ophthalmology
Orthopedics
Orthotics
Pain
Physiotherapy
Plastic Surgery
Psychiatry

Team Responsibilities

Dermatology Physicians
Your child's dermatology physicians are the primary physicians responsible for the overall plan of your child's care. He or she will discuss your child's medical condition and treatment with you, including ways to protect your child's skin from blistering, preventing infection, minimizing deformities and monitoring of possible complications associated with EB. He or she will liaison with other members of your child's healthcare team, and address any of your medical concerns and questions.

Dermatology Clinic Nurse
Your child's clinic nurse is the person who can answer your questions and give you the support and advice you need in regards to your child's daily care; including wound care and home care. He or she will work directly with you and your child's primary physician and EB team to plan and coordinate your child's care. Your clinic nurse has expert knowledge about your child's disease, treatment, and its problems. When you are at home and have questions or concerns, you can call or email your clinic nurse.

Dermatology Social Worker
Your social worker is there to look out for the social and emotional needs of both your child and your family. He or she can help you deal with the social, emotional, and financial aspects of your child's illness and treatment. He or she can also give you information about finding hospital services, community groups, and groups that provide benefits that can be useful for your child's illness.

Dermatology Information Coordinator
Your child's healthcare team will also include an Information Coordinator, who books and coordinates appointments, tests and procedure that the other members of your child's healthcare team may ask for.

Dermatology Clinic Clerk
He or she will register your child when you first arrive at the clinic and will schedule your next clinic appointment. If you need to reschedule an appointment because you can't come to clinic, this is the person you need to contact and inform of the cancellation and she will help you reschedule the appointment.

Dermatology Fellows
A dermatology fellow is either a paediatrician or dermatologist that is completing further training in paediatric dermatology. Your child may be seen by different fellows at clinic appointments. They are in training, so a staff physician will also see you during your visit.

Dermatology or Paediatric Residents
A resident is a physician who is training to be a general paediatrician or dermatologist. You may also be seen by a resident, and because they are also completing further training the staff physician will again see you as well.

As one of the teaching hospitals for the University of Toronto, SickKids trains people from around the world to become doctors, nurses, and other health-care workers. During your visit, your child may be seen by one of these students. Be assured that these students are well-supervised by our professional staff.

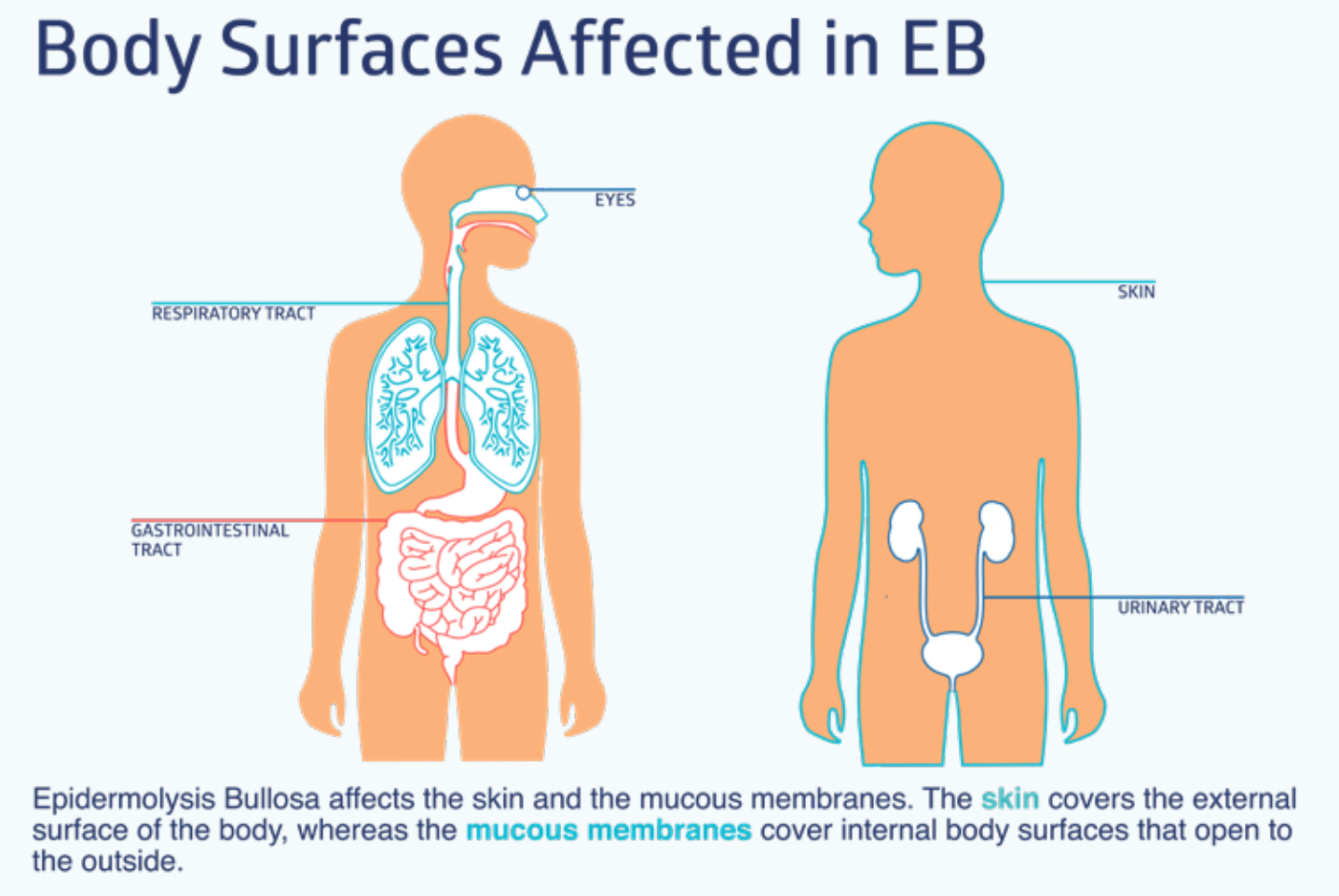
What is EB?

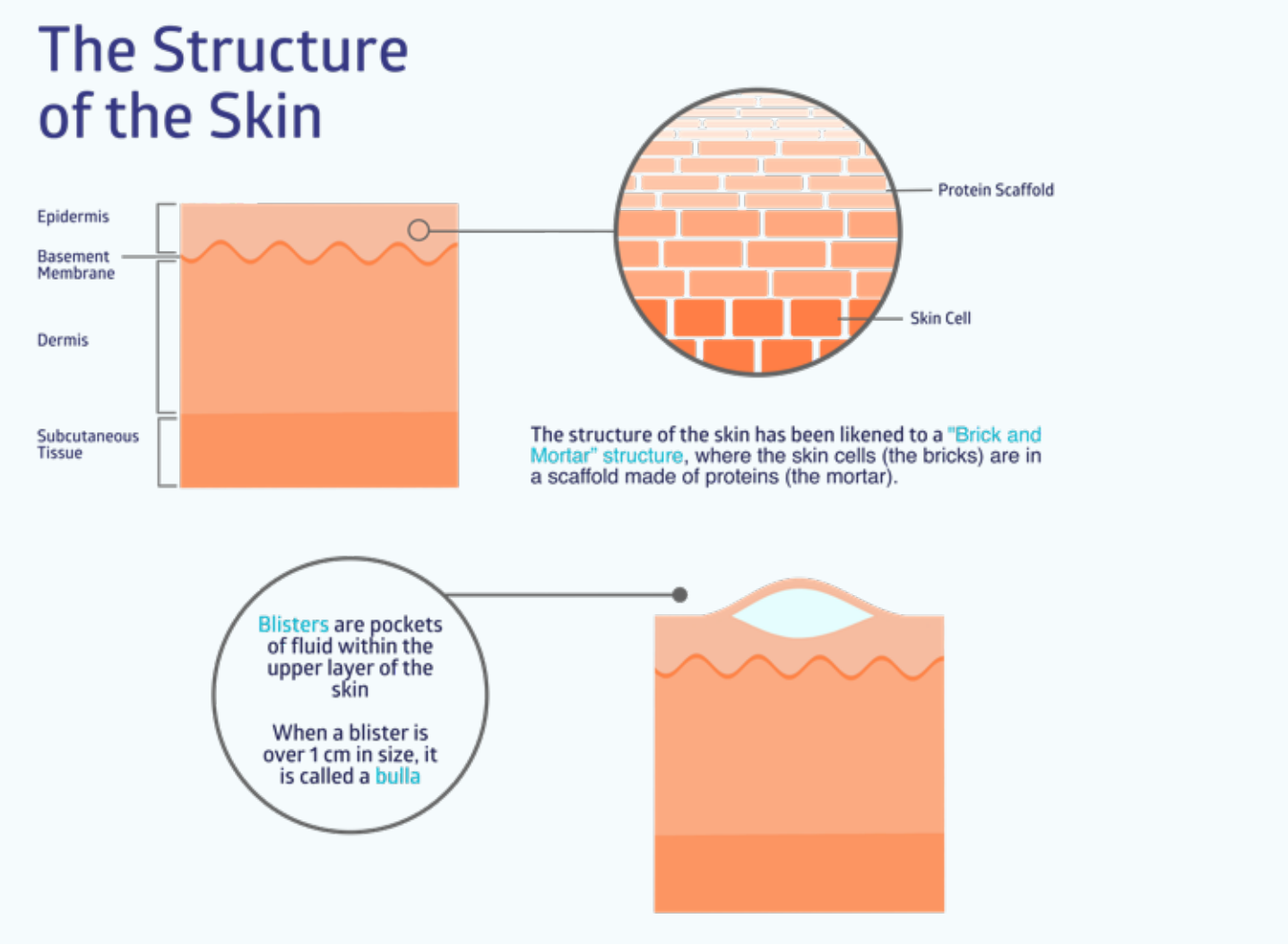
Elena Pope, MD, MSc, FRCP(C)

Epidermolysis Bullosa (EB) is a group of skin diseases that cause various degrees of skin and mucous membrane fragility. The skin becomes fragile when proteins essential for skin integrity are absent, reduced, or abnormal.

The skin can be described as a brick wall, where the skin cells ("the bricks") are in a scaffold made of proteins ("the mortar"). Even one abnormal protein may weaken the skin scaffold. The weakened scaffold then results in the separation of the skin layers and accumulation of fluid within the skin, called a **blister**. The main skin finding in all types of EB is a fluid- or blood-filled blister, or bulla (a blister that is > 1 cm in size). In contrast with other skin conditions that cause blisters (for example, burns, infections, etc.), EB causes life-long skin fragility and blistering.

Over 16 proteins have been reported as a cause for EB. For that reason, clinical presentation of the disease is quite variable. The severity of skin and other organ involvement depends on the degree of protein abnormality (absent, reduced, abnormal, etc.) and the location of the abnormal protein in the skin layers. Deeper locations are typically associated with more severe clinical presentations.





What are the Different Types of EB?

Currently, EB is classified into four different subtypes, depending on the location of the abnormal or missing protein in the skin: Epidermolysis Bullosa Simplex (EBS), Junctional Epidermolysis Bullosa (JEB), Dystrophic Epidermolysis Bullosa (DEB), and Kindler Syndrome.

Epidermolysis Bullosa Simplex (EBS)

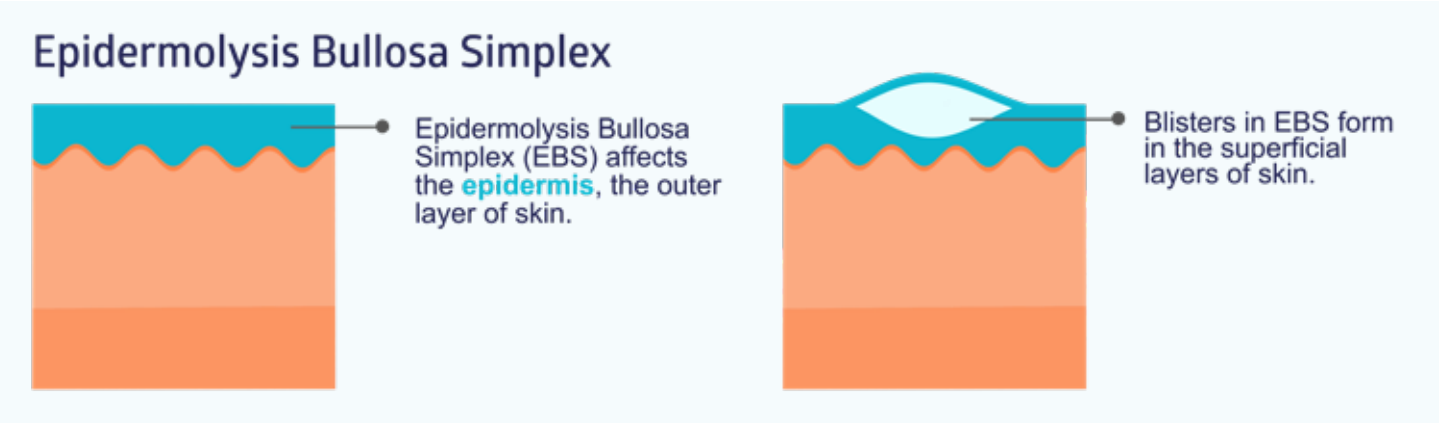
EB simplex (EBS) is caused by abnormalities in proteins present in the outer layer of the skin, the epidermis. These proteins are called keratins (genes *KRT5* or *KRT14*). There are several subtypes even within this group, accounting for various degrees of blistering. Typically though, patients with EBS have milder disease since the proteins involved are higher up in the skin. In most cases, but not always, there is a family history of similar blistering in first-degree relatives (parents, siblings).

Classical presentation of EBS includes:

- Blisters, primarily of the palms and soles
- Blistering is worsened or provoked by trauma (for example, shoes, gardening, etc.) and heat

- Blisters heal over a few days with no permanent scarring
- No or very mild mucosal involvement (for example, no mouth blisters)
- Changes in the nails are rare

A rare type of EBS, called **Dowling Meara**, has more skin involvement with blisters that expand at the edges causing a “string of pearl” appearance, as well as more significant palm and sole involvement causing thickening of the skin (keratoderma). This may result in difficulty with walking or using hands. Blistering can be more extensive shortly after birth in the generalized forms of EBS and in EBS caused by rarer abnormalities in structures other than keratins (desmosomal genes *PKP1* or *DSP1*).
Other forms may present only when the chances for friction increase (child starts crawling or walking).



Junctional Epidermolysis Bullosa (JEB)

Junctional EB (JEB) is caused by abnormalities in proteins found at the meeting point (basement membrane) between superficial (epidermis) and deeper skin layers (dermis). Abnormalities in several genes may cause JEB. All these genes are inherited in an autosomal recessive manner. This means that both parents are carriers of the abnormal gene but do not have any manifestation of EB, and one needs two abnormal copies of the gene to have the disease. There are two major clinical subtypes of JEB:

1. Herlitz JEB

Herlitz JEB is caused by abnormalities in three genes encoding a protein called laminin 332 (genes *LAMA3, LAMB3, LAMC2*). It is one of the most severe subtypes of EB and is characterized by:

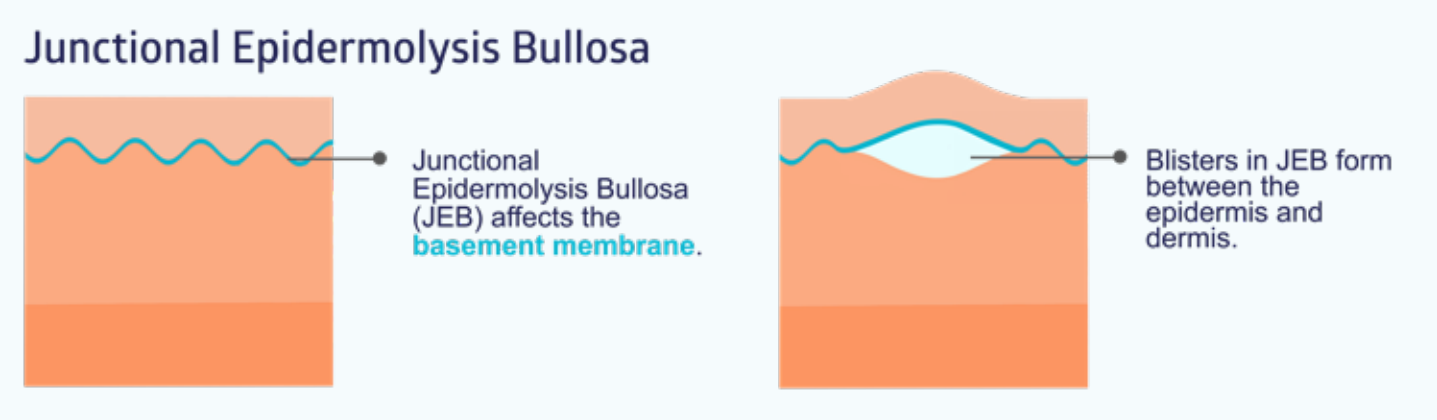
- Severe skin fragility resulting in widespread blistering and erosions
- Mucosal fragility leading to airway involvement (hoarseness, trouble breathing, etc.)
- Typical presentation with blood tinged blisters and/or crusting around the tip of the nails
- Blistering and crusting around the mouth, eyes, nose, etc.
- Blisters/erosions may take a long time to heal creating a heaped up (“crater-like”), fragile healing tissue that bleeds easily
- Increased need for calories that cannot be met via oral feeds leading to poor growth
- Chronic anemia and low protein levels
- High risk for infection and sepsis (severe infection causing injury to tissues and organs)
- High risk for sudden breathing difficulties from new blister formation
- Limited life-span; most children succumb due to complications in the first few years of life

2. Non-Herlitz JEB

Non-Herlitz JEB may be caused by abnormalities in the laminin protein and additionally from abnormalities in the type XVII collagen protein (gene *COL17A1*), abnormalities in the α6 or β4 integrin proteins (genes *ITGA6* or *ITGB4)* or very rarely, abnormalities in the plectin protein.

- Similar presentation with the Herlitz variant, but of lesser severity
- Dental enamel defects causing dental caries
- Nail involvement
- Patchy scarring hair loss (alopecia)
- Eye involvement
- Survival into adulthood

A very rare form of JEB can present shortly after birth with vomiting/feeding intolerance due to an absent pylorus (a muscular ring at the junction between stomach and the first part of the intestine). Patients who are promptly recognized and surgically corrected will do well from gastrointestinal point of view, but may continue to present various degrees of blistering throughout their lives.



Dystrophic EB (DEB)

Dystrophic EB (DEB) is caused by abnormalities in the type VII collagen protein (gene *COL7A1*). This form of EB may be inherited in either an autosomal dominant or autosomal recessive manner. Type VII collagen is essential for structures called anchoring fibrils that, as their name suggests, attaches the superficial layer of the skin to the deep layer of the skin. As it is deeper in the skin, blisters are also deeper, take longer to heal and may leave scars after healing. The location of the blisters is variable from patient to patient but tends to affect pressure and trauma-prone areas.

1. Recessive DEB (RDEB)

Patients have different clinical presentations, but most have involvement of the skin, mucosal membranes with various degrees of complications.

Skin involvement:

- Deep blisters, longer healing time, healing with scarring
- Difficult to heal wounds (chronic wounds) causing significant degree of pain
- High risk of skin colonization or infection with bacteria, fungi
- Large areas of the skin involved

Mucosal involvement

- Eye involvement presenting as painful erosions; may lead to scarring and blindness if not treated
- Mouth involvement with painful erosions causing difficulty eating and performing dental hygiene; overtime may cause limitation in mouth opening and stiffening of the tongue
- Gastrointestinal tract involvement; blistering of the esophagus (feeding tube) that may lead in time to narrowing (strictures); gastro-esophageal reflux; constipation
- Urogenital tract involvement: kidney infections, strictures of the urinary tract, potential to renal failure

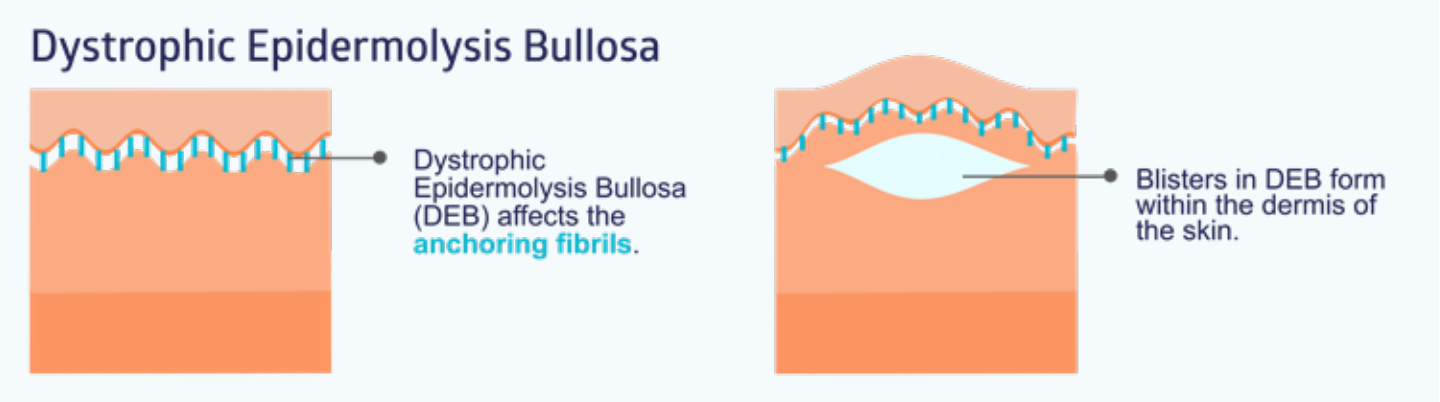
Other system/organ involvement

- Anemia
- Low protein levels
- Potential for enlarged heart
- Weak bones (low bone density)
- Poor growth and development

Patients with RDEB have a high risk of infection and complications related to their skin and other organ involvement. Their life expectancy is limited to early adulthood; however, this is highly variable. Most patients surviving into adulthood may develop aggressive skin cancers called squamous cell carcinoma (SCC).

2. Dominant DEB (DDEB)

DDEB has a milder presentation than RDEB. Typically, patients have a more limited skin involvement. Blisters develop in high friction areas that heal with scarring and milia (white, small and hard papules). Nail involvement with nail dystrophy or absent nails are very common. Although the potential for complications exists, similar to RDEB, complications are not common in DDEB.



Kindler syndrome

Kindler syndrome (KS) is a rare inherited skin condition that also presenting with skin fragility early in life. It is inherited in an autosomal recessive manner. These children develop blisters at different levels of the skin, so some blisters might heal without scarring, while others might heal leaving scars behind. Overtime, the blistering and skin fragility improve, but these are replaced with more scarring and hyperpigmentation. There is increased sun sensitivity that further contributes to skin damage, leading to scarring, hyperpigmentation (darker pigment skin) and areas of skin thinning (atrophy). Patients with Kindler syndrome have significant gum inflammation.

Making the Diagnosis

Irene Lara-Corrales, MD, MSc

Making a diagnosis of EB, particularly the subtype of EB, might not be easy and takes time. It is important to recognize that skin fragility in newborns is encountered in conditions other than EB. For example, there are other genetic and infectious diseases that can present with blisters and fragile skin early in life. Preterm babies as well have very fragile skin due to prematurity and don't experience blistering later in life. Although we do need to think of EB if we see blisters developing in a newborn, it is important to have in mind that very early on we are unable to confirm a diagnosis just by looking at the baby's skin.

How do we make a diagnosis of EB?

There are several ways to diagnose EB. If other family members are affected by EB and a baby has blisters at birth or early in life, it is very likely that the same diagnosis is present in the baby. In the absence of a positive family history, we will be suspicious of the diagnosis when we see blisters at birth or developing in areas of friction or trauma.

How can we be sure of the diagnosis?

There are two different ways of confirming the diagnosis:

Skin Biopsy

Sometimes we rely on skin biopsies to give us more information. A skin biopsy is a small piece of the skin that is sent to special doctors that look under the microscope and are able to provide more information about the blister: how deep it is, if the structures of the skin look normal or abnormal, and if there are cells or other changes that help us make a diagnosis. The blisters in EB occur at different levels depending on the type of EB; a biopsy can sometimes tell us the level of the blister in the skin and help us determine the type of EB we are dealing with. There are special tests as well that help us look at the different proteins that might be affected in EB – these tests are very useful for helping us make a diagnosis.

Genetic testing

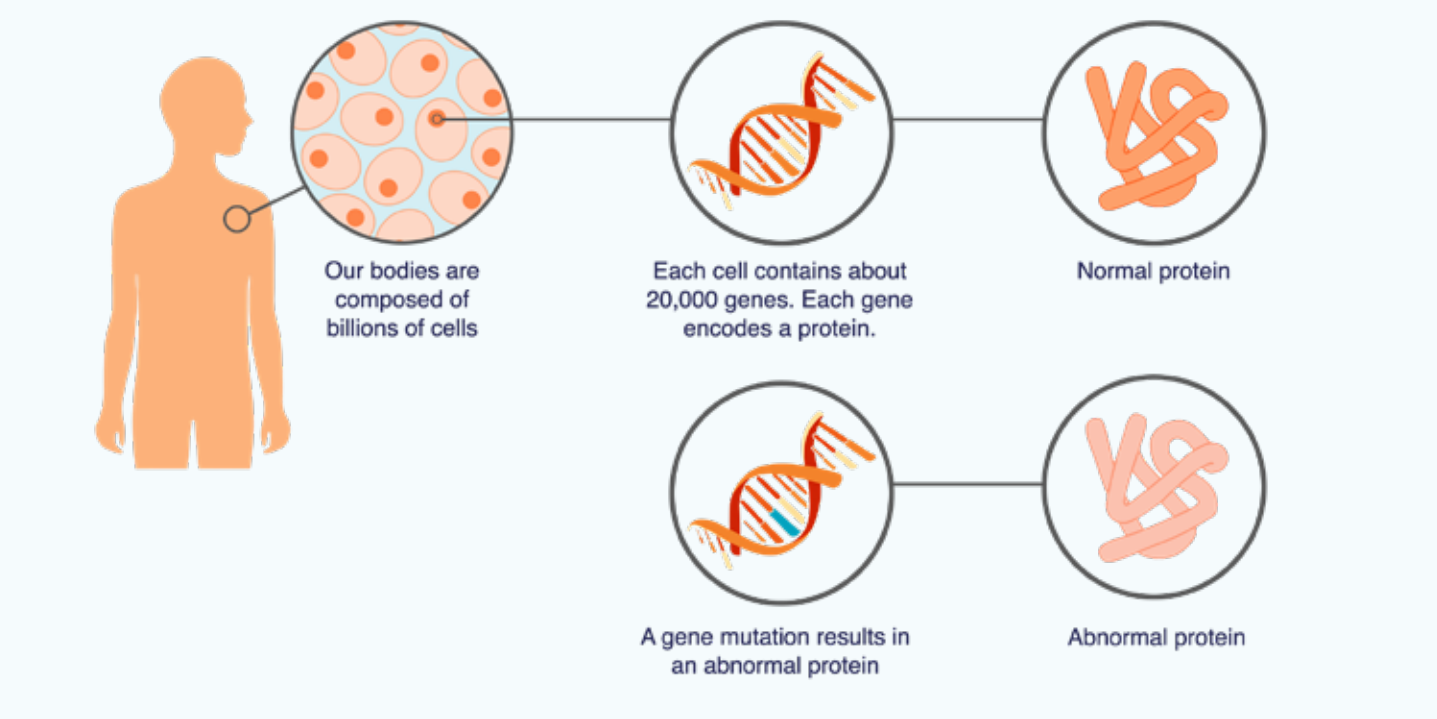
Although skin biopsies are useful, they might not always give us an answer. Sometimes we need other tests to confirm the diagnosis. Genetic testing uses a blood sample from the patient to give us more detailed and definitive information. Because EB is a genetic disorder, we are able to look in the blood of patients to identify changes in the genetic material that could lead to EB. If we find changes in proteins that are associated with EB, we are able to confirm the diagnosis and also know the exact type of EB we are dealing with. This is useful because we can give families more information about the type of EB the patient has.

The Genetics of EB

Peter Kannu, MD, PhD and Andrea Shugar, MS, CGC

What are genes?

Our bodies are made up of billions of cells. **Genes** are the instructions that our cells use to make proteins. Each **protein** has a unique and specialized job in the cell. Everyone has two copies of each of their genes. One copy of each gene comes from the mother (through her egg) and the other comes from the father (through his sperm). In this way, both parents contribute equal numbers of genes to their children.



Which genes are responsible for EB?

There are more than 17 different genes that are known to cause several different forms of EB when defective. All of these genes play a role in the formation of the epidermis (the outer layer of the skin). When an EB gene stops working properly, the protein it encodes is not made properly. Genes stop working properly when they develop errors (mutations) in their code (just like a computer program that stops working when it has an error in its code).

Genetic testing is available for many of these mutations. The sensitivity of genetic testing is very high (99%). This means that a person with EB who undergoes genetic testing has a very high chance of finding out which gene caused their condition. In a very small number of people with EB (less than 1%), we are unable to identify the specific genetic cause (gene).

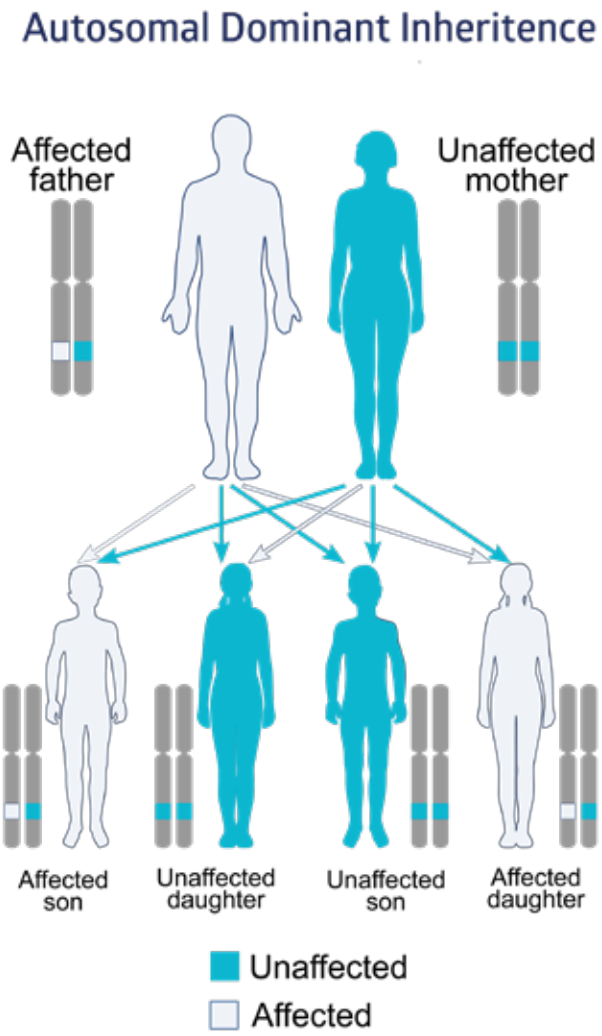
How is EB Inherited?

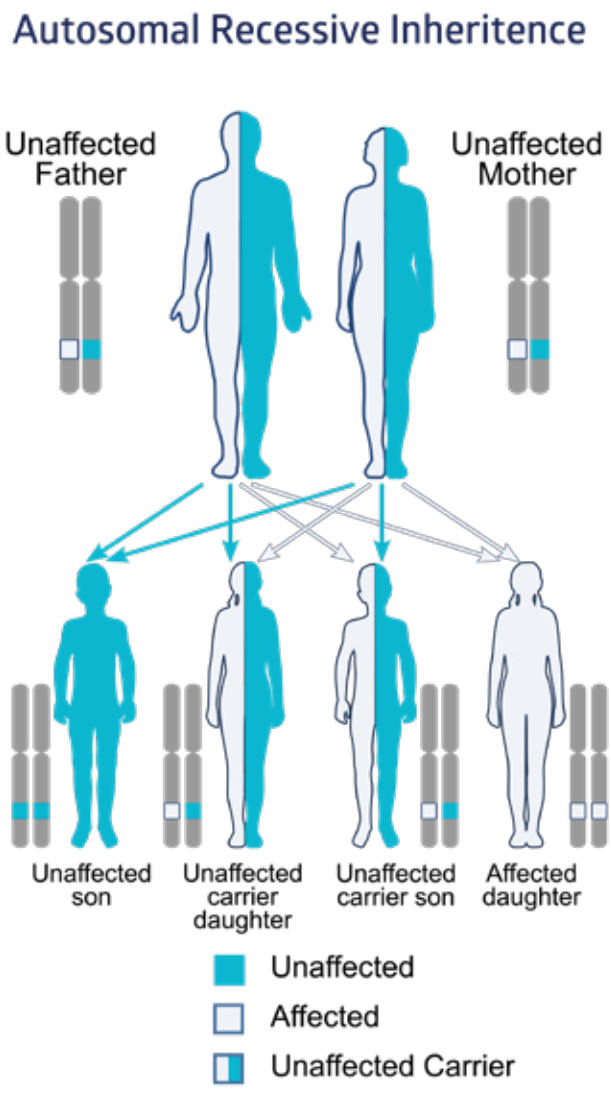
Most forms of EB are inherited in an **Autosomal Dominant** manner:

- People with an autosomal dominant form of EB have a mutation in only one of their two EB genes. One mutation is sufficient to cause the condition.
- Autosomal dominant conditions affect males and females equally
- A person with dominant EB has a 50% (1 in 2) chance to have a child with EB in each pregnancy they have with their partner.
- A person with dominant EB also has a 50% chance to have a child without EB in each pregnancy they have with their partner

Some forms of EB are inherited in an **Autosomal Recessive** manner:

- People with an autosomal recessive form of EB have a mutation in both copies of their EB gene
- Autosomal recessive conditions affect males and females equally
- Parents of affected children are “carriers” of a single mutation, but do not have EB
- Two carrier parents have a 25% (1 in 4) chance to have an affected child in each pregnancy they have
- Two carrier parents also have a 75% to have an unaffected child in each pregnancy they have





How can genetic testing and genetic counselling help?

Genetic testing may help you to:

- Identify the specific gene and mutation that caused the EB in your family
- Pinpoint the exact type of EB that is in your family
- Help to identify other carriers of EB who would be at increased risk to have an affected child

Genetic counselling may help you to:

- Better understand the EB diagnosis in your family and how it is inherited
- Identify who might be at risk to be a carrier or to be affected in your family
- Explore your feelings related to the diagnosis and connect you with appropriate resources.
- Understand your options for future family planning, including preimplantation genetic diagnosis (PGD) and prenatal diagnosis (PND)

How can I access genetic counselling or genetic testing?

Ask your healthcare provider to refer you to your local genetics clinic. You can find a genetics clinic near you by visiting the Canadian Association of Genetic Counsellors (CAGC) Clinic Search Page: https://www.cagc-accg.ca/?page=225

Newborn with EB

Irene Lara-Corrales, MD, MSc and Michelle Lee, RN, BScN

Feeding

Good nutrition is especially important in children with EB, not only for their healthy growth and development, but also for wound healing. Some children with EB may have blistering in the mouth, which causes difficulties with feeding. Most babies with EB cannot breastfeed properly, so expressed breast milk is recommended. It is important to use a soft nipple, to reduce the amount of blistering in your child's mouth. A Habermann nipple (available at the SickKids specialty food store), Mead Johnson Cleft Palate Nurses, or the Pigeon feeder (www.cleftadvocate.org) can be used to avoid oral blisters. The Habermann nipple and the Mead Johnson Cleft Palate Nurses are X-cut nipples, which allow the milk to flow only when your child sucks on it. This reduces the amount of air your child swallows. The Pigeon feeder has a Y-cut nipple, which does not collapse when your child sucks, and fits naturally in your child's mouth.

The Habermann Nipple

The Mead Johnson Cleft Palate Nurse

The Pigeon Feeder

For easier sucking, moisten the nipple or soother with water. If you do choose to breastfeed your baby, apply Vaseline to the nipples and on your baby's face to reduce the friction.

For your child's oral health, gently clean your child's mouth with a spongy toothette. As long as your child is eating well, blisters in the mouth can be left alone, because sucking and chewing will cause the blisters to puncture and drain.

If your child's weight gain is poor, you should tell your Primary Physician, who may refer you to the Gastroenterology (GI) Clinic. Here, your child may be seen by a dietitian, who can help you develop a nutritional plan that is best for your child's growth and that is easy for him or her to chew and digest. If your child is unable to eat by mouth, your doctor may recommend a feeding tube to deliver food directly into your child's stomach.

Clothing

Dress your child in loose-fitting clothing made of soft, breathable fabric such as satins, silks, and soft cottons. Choose seamless clothing, or turn clothing with seams inside out so that the seams are facing outwards. Avoid clothing that have tight elastic bands, harsh buttons, snaps and zippers, and remove clothing tags and labels before use. Wearing soft undergarments or bandages may be necessary to protect the skin. Keep in mind when choosing clothing that it must be easily put on and taken off. Take special note of head, ears, and armpit areas when changing your child.

For infants, placing cotton mittens over their lubricated hands and feet can prevent them from scratching or rubbing the face. To protect elbows, you can take a large pair of socks and cut the toes off, and then pull the socks over the elbows to protect the elbows from rubbing. If your child gets blisters from the sides of the crib, you can pad elbows, heels and knees with gauze sponges secured by rolled gauze. Full sleepers, which include feet, are recommended. For young children who are learning to crawl and walk, consider adding foam padding to the clothing in the knee and elbow area.

Recommended Products

Product Name	Description	Provider
BabyLegs	Like legwarmers, to hold bandages in place on arms and legs	BabyLegs www.babylegs.net
Tubifast Garments	Garments for dressing retention	Monlylcke www.molnlycke.com
Bees Knees Crawling Pants	Pants with foam cushions in the knee area	Bees Knees www.beeskneesbaby.com

Bedding and Car Seats

For extra padding, you may choose to add sheepskin or egg crate foam to your child's bed or crib, car seat, and seat belt. Do not lay your child directly over the sheepskin or foam. Jersey-knit or satin sheets are suitable for use by children with EB. With satin sheets, keep in mind that if blood or other fluid leaks and dries on the sheet, it will become hard and this can cause blistering.

Wound Care in EB

Irene Lara-Corrales, MD, MSc and Michelle Lee, RN, BScN

Blister Management

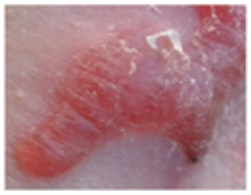
Although you should take precautions to minimize the formation of blisters, it is impossible to completely prevent your child's skin from blistering. When your child does develop blisters, they need to be drained or lanced to prevent the blister from growing and creating a larger wound.

Before lancing blisters:

- Make sure the area you are working in is clean.
- Wash your hands thoroughly.
- Make sure your tools are organized and cleaned or sanitized:
  - Sterile needle, lancet or scissors specifically for lancing blisters
  - Sharps container for used needles (thick plastic or glass container that you are able to close). You can dispose of container at a local pharmacy
  - Gauze to wick away fluid
  - Towel to provide clean area around skin and tools
- Pain control medication if required: to be given 20-30 minutes prior to beginning
- Distraction: toys, music, TV, electronic devices

Lancing Blisters Step-by-Step Approach:

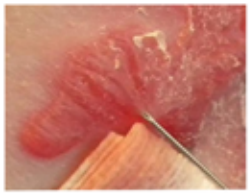
1



Aim for the edge of the blister.

If you are using a needle or lancet, do not aim directly at your child; instead, aim parallel to the skin.

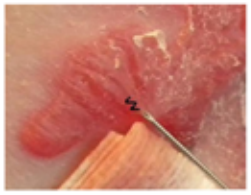
2



Very carefully, puncture the blister with the needle, lancet or scissors.

If you are using scissors, you can pinch a small section of the blister cap between the scissor blades and create a small slit.

3



Depending on the size of blister, you may need to puncture multiple areas to make sure that the opening is large enough for the fluid to drain and to make sure the blister will not reseal.

Let fluid drain from blister; you may use sterile gauze to absorb fluid if necessary.

Do not apply pressure on the top of the blister.

4

Leave the overlying skin in place

Repeat this procedure as soon as your child develops new blisters. It is recommended that you check your child's skin daily.

5 **Protection**

If the lanced blister is large or still draining, silicone dressings or non-adherent contact layers are recommended because they don't stick to the skin.

Depending on the location of the blister, certain dressings may work better than others. Dressings should be kept on for a couple days, but if you suspect that there may be fluid coming from the wound, daily dressing changes should be done to reduce the risk of infection. Topical antibiotics are not necessary for clean, non-infected wounds.

Dressing Supplies

Tools

You will need sharp scissors to cut and trim bandages. Bandaging scissors are available from most medical supply stores. Fine scissors, such as manicure scissors from your local drugstore can be used to pop blisters and trim dead skin. Clean your scissors before and after use by washing them with soap and water, or wiping them with rubbing alcohol. Needles, lancets or scalpels can also be used to pop blisters, and they are available from medical supply companies and some pharmacies.

Ointments

Aquaphor Healing Ointment or Vaseline are highly recommended, both of which are available at most drugstores without a prescription. Talk to your child's doctor before using any ointment with active ingredients, such as anti-itching creams. Do not use highly-sensitizing antibiotics, such as Bacitracin, without first consulting your child's doctor.

Dressings

The following table contains recommendations of supplies that have been used by EB families, to help you get started. There are many options and brands available that can be used for your child's care, and you may choose to experiment with different types to see which products work best for your child.

Dressing Type	Product Name	Picture	Provider	Use in EB
Foams: Silicone	Mepilex		Molnlycke Health Care www.molnlycke.com	For wounds in areas that may be easily rubbed/hit; provides extra padding and protection
	Mepilex Border			
	Mepilex Border Lite			
Foams: Hydrogels	Duoderm gel		Convatec www.convatec.ca	For wounds that have dry and crusty areas in need of moisture
	IntraSite gel		Smith & Nephew www.smith-nephew.com	
Foams: Alginates	Aquacel		Convatec www.convatec.ca	For wounds that have a lot of fluid coming from the wound: *Requires frequent dressing changes* *Not suitable for wounds that have little or no fluid*
	Kaltostat		Convatec www.convatec.ca	
	SeaSorb		Coloplast www.coloplast.ca	
Hydrocolloids	Restore		Hollister www.hollister.com/canada	Should not be used for EB as it is adhesive
Others	Telfa		Kendall Company Ltd www.kendallhq.com	For wounds that may have some fluid coming from the wound

Dressing Type	Product Name	Picture	Provider	Use in EB
Silicone Tape	Mepitac		Molnlycke Health Care www.molnlycke.com	To tape down dressings; suitable for EB skin
Silicone Mesh	Mepitel		Molnlycke Health Care www.molnlycke.com	For wounds that require topical medications: *Requires a secondary dressing (e.g. Telfa) Can be washed with tap-water and reused*
Silver Dressings	Aquacel Ag		Convatec www.convatec.ca	For critical wounds or wounds that are infected
	Contreet		Coloplast www.coloplast.ca	
	Mepilex AG		Molnlycke Health Care www.molnlycke.com	
	Acticoat		Smith & Nephew www.smith-nephew.com	
	Silvasorb		Medline Industries www.medline.com	
	Restore AG		Hollister www.hollister.com/canada	
Gauze	Vaseline Petrolatum Gauze			Inexpensive option, but will stick to the skin when it becomes dry and requires more frequent dressing changes
Burn net or cling				Holds dressings in place

In addition to your local drugstores, special dressings can be purchased from the following medical providers:

Medical Mart Home Health Care Store
550 Matheson Blvd W, Unit 101
Mississauga, ON L5R 4B8
905-624-2011
www.medimart.com

Starkmans Health Care Depot
1243 Bathurst Street
Toronto, ON M5R 3H3
416-534-8411
www.starkmanshealth.com

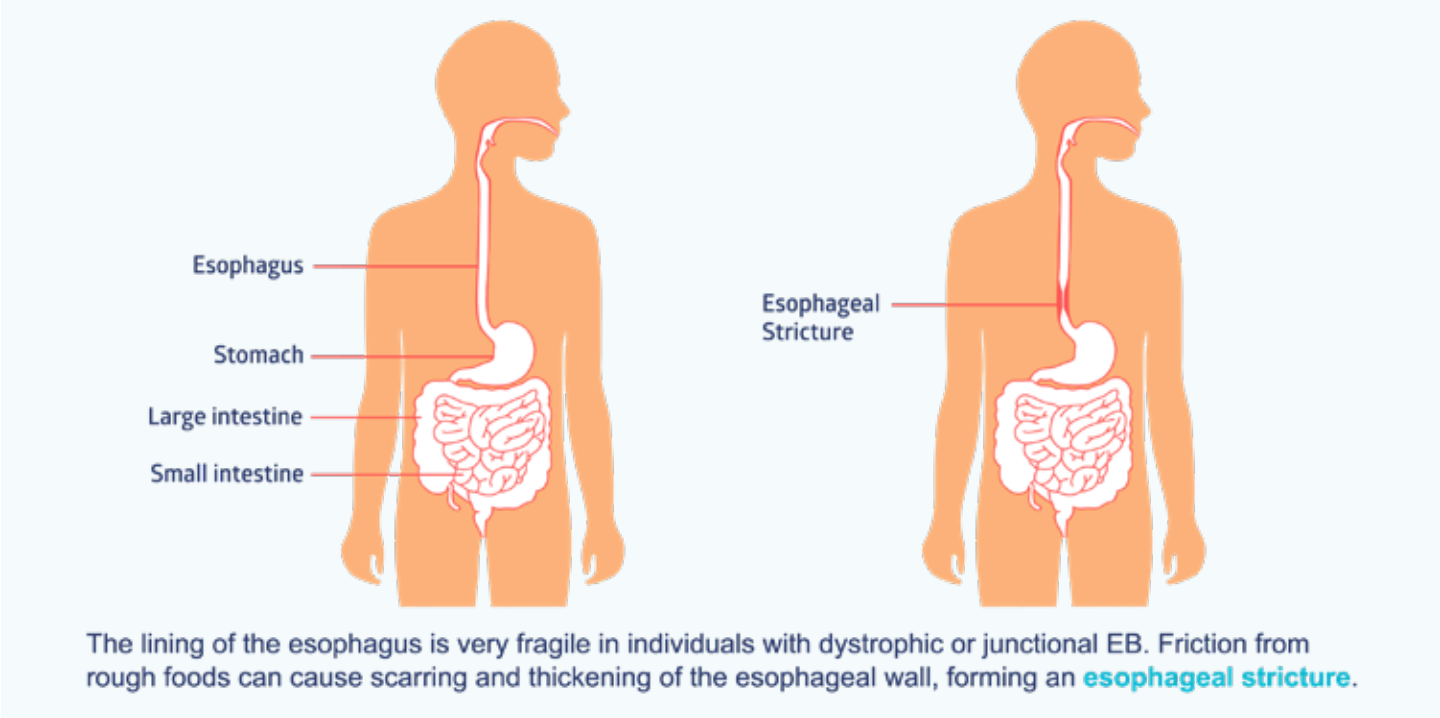
Calea
905-624-1234
www.calea.ca
Shoppers Drug Mart Home Health Care
Visit their website for the location of the store closest to you
www.shoppershomehealthcare.ca

Esophageal Strictures in EB

Margaret Marcon, MD, FRCP(C) and Carmen Liy-Wong, MD

What is an Esophageal Stricture?

An **esophageal stricture** is a narrowing in the esophagus, the muscular tube that carries food and liquids from the mouth to the stomach. It is the most common gastrointestinal complication in recessive dystrophic and junctional EB. The narrowed esophagus makes it difficult to swallow food and sometimes even liquids. This complication is a major cause of poor nutrition in recessive dystrophic and junctional EB. Not only does a stricture in the esophagus affect the intake of nutrients, it also limits food choice. The patient's favorite foods are often removed from their diet, affecting the simple enjoyment of eating, an important part of family and social life. This makes esophageal strictures one of the most debilitating features of EB.



The lining of the esophagus is very fragile in individuals with dystrophic or junctional EB. Friction from rough foods can cause scarring and thickening of the esophageal wall, forming an **esophageal stricture**.

How Does an Esophageal Stricture Form?

The esophagus in individuals with dystrophic and junctional EB has extremely fragile surface lining that makes it easy to blister in response to even the most minor trauma (hot or rough foods). The blistering can then lead to the formation of scar tissue in the wall of the esophagus, and can cause it to narrow or even get blocked. Esophageal strictures usually begin in childhood, with more than half of those with dystrophic or junctional EB reporting symptoms by 10 years of age. The risk then increases as one gets older.

What are the Symptoms of an Esophageal Stricture?

- Difficulty swallowing (dysphagia)
- Pain with swallowing
- Weight loss or difficulty gaining weight and poor growth
- Regurgitation of food, when food comes back into the mouth from above the stricture
- Food gets stuck in the esophagus (food impaction)
- Frequent burping or hiccups
- Heartburn (burning sensation behind the breast plate bone)

If your child is having any difficulty swallowing, taking much longer to eat or having heartburn, mention this to their health care provider. They will ask you about their symptoms and decide if further testing is necessary.

Tests may include:

- **Barium swallow test**: For this test you swallow liquid barium, which coats and fills the esophagus, so that it shows up on X-ray images. X-ray pictures are then taken and the radiologist can see if there is a narrowing in the esophagus. Barium is nontoxic and is often flavored to improve the taste.
- **Upper gastrointestinal endoscopy**: Your doctor will place an endoscope (a flexible tube with a light and video camera attached) through your mouth and into your esophagus. It allows your doctor to examine your esophagus and upper intestinal tract.

How are Esophageal Strictures Treated?

- If there is a significant stricture, a dilatation (stretching of the stricture) will be most often be performed. **Esophageal dilatation** is a procedure where a dilating device, often a balloon, is placed across the stricture and then inflated to stretch the stricture open and widen the esophagus at that point. The dilatation can be done through the mouth by placing the balloon under X-ray guidance or through an endoscope under the guidance of a camera. For comfort, this procedure may be performed under sedation, including a general anesthetic. A local anesthetic spray may also be applied to the back of the throat. Repeat dilatations are often required to adequately stretch the esophagus. Because EB is a lifetime condition, multiple procedures are often required because of recurrence.
- Despite esophageal dilation, some patients present with significant failure to thrive and require the placement of a **gastrostomy tube** (also called G-tube) is a tube inserted through the abdomen in the stomach. The tube delivers nutrition directly to the stomach.
- Sometimes, when esophageal strictures are worsened by acid reflux from the stomach into the esophagus, medications such as acid blocking drugs (proton pump inhibitors or H2 blockers) may be added to prevent the stricture from returning. Corticosteroids may also be added and can be given in a form that coats the esophagus (topical) or that is absorbed in the stomach (prednisone pill/liquid).
- Sometimes, extra calories are given by the intravenous route to supplement your child's nutrition. If there is a G-tube (gastrostomy) in place, the dietitian may increase the calories given via this route.

Malnutrition in EB

Inez Martincevic, MSc, RD and Margaret Marcon, MD, FRCP(C)

Individuals with severe EB, especially those with recessive dystrophic EB or junctional EB, are at risk of malnutrition and require nutritional support. There are several reasons for poor nutrition in a child with EB:

- Eating challenges, including:
  - Mouth sores
  - Reduced ability to self-feed
  - Poor appetite (does not feel hunger)
- Fragile mucosal lining of the gastrointestinal tract "gut", which causes:
  - Ulcers, blistering, and pain
  - Difficulty swallowing
  - Esophageal strictures
  - Reduced absorption of nutrients
- Increased metabolic rate and energy requirements due to:
  - Excessive loss of heat and nutrients through the fragile skin
  - High skin turnover and wound healing
  - Frequent infection
  - Inflammation (red, sore and swollen tissue)

It is important to provide nutritional support, as malnutrition is associated with:

- Poor growth
- Delayed puberty
- Poor bone health
- Fatigue
- Limited ability to be physically active
- Potential for poor quality of life

It is important for children with EB to be evaluated regularly by a dietician in order to assess their nutritional requirements and provide an individualized nutrition plan.

Goals of Nutritional Support

- Optimize growth
- Promote puberty
- Correct nutrient deficiencies
- Alleviate stress with eating
- Optimize healing
- Improve bowel function
- Optimize immune health

Nutritional Support in EB

General nutrition recommendations for children with EB include:

Breast Feeding Babies

If breast-feeding is not possible or on its own is not enough for weight gain, supplementing with formula may be required.

Growth

In cases where growth is not ideal (this means the weight or height curves are flat or a child has dropped on a curve) then supplementing with formula either by mouth or using a tube may be advised. Tubes can go through the nose, a nasogastric (NG) tube, and are short term plans. Tubes can also go through the stomach, a gastrostomy (G) tube, and are for long term nutrition support, for example sometimes in children with severe dystrophic EB.

Nutritional Deficiencies

Regular monitoring of nutrient levels, including iron, calcium, vitamin D, phosphate, vitamin A, vitamin E, zinc, selenium, B vitamins like folate and vitamin B12, urea and carnitine

Constipation

It is common for children with EB to have problems passing stool. Fluids are very important to help pass stool, especially if taking medications to soften stools.

Assessing and Treating Pain caused by EB

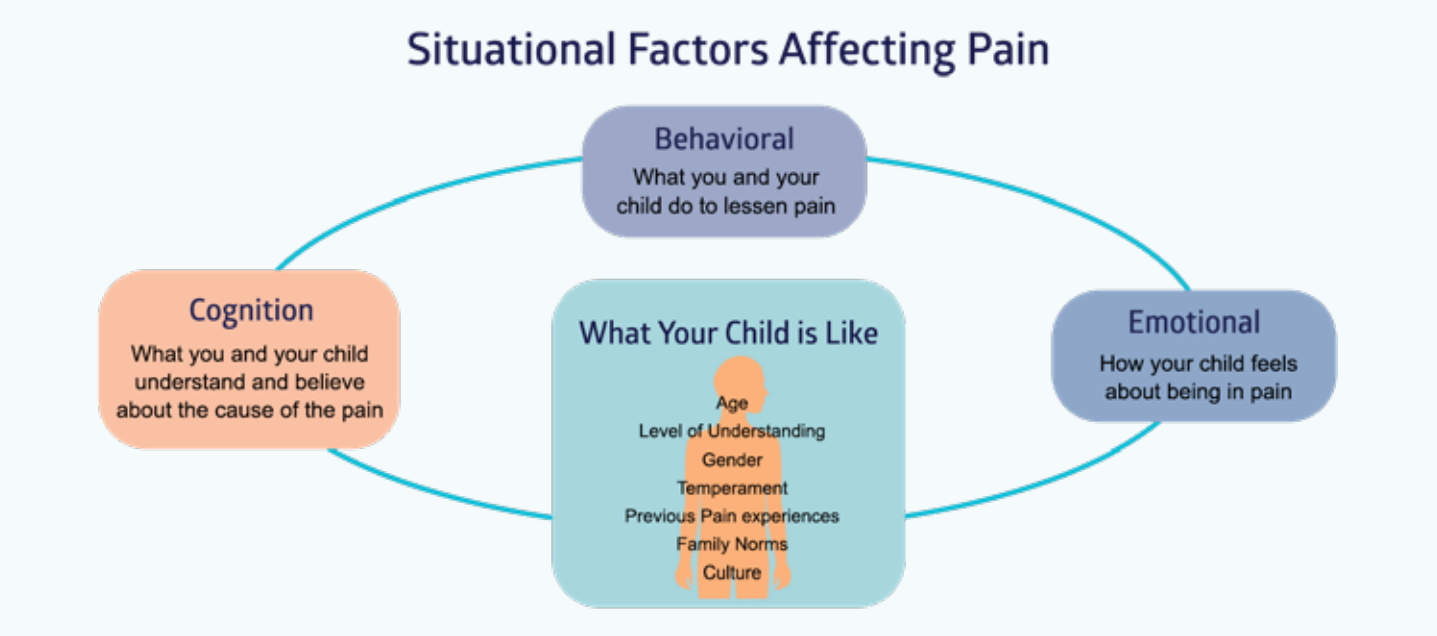
Fiona Campbell, BSc, MD and Jennifer Tyrrell, RN, MN

Understanding Pain

Acute pain is an unpleasant sensation that serves as an alarm, designed to protect us from injury. Pain is one of many self-protective mechanisms of our body. It exists so that we can quickly take steps to remove the source of the injury. Pain may also alert us so that if a part of the body hurts, we can take steps to help it heal, such as resting or having it treated. Thus, Acute pain tells us to pay attention and get treatment.

Chronic pain is pain that lasts longer than three months and can be continuous (experienced all the time) or intermittent (comes and goes). It can sometimes be the result of changes in the sensitivity of your pain nerves, which produces the feeling of pain even when there is no injury or danger. In some people, the nervous system becomes "sensitized" to the pain message after an injury; this means that the pain message continues to be sent to our brain even after the tissues have healed, like a false alarm. Chronic pain can often affect mood, sleep and daily activities, such as attending school, participating in hobbies and playing with friends.

We know from experience and research that pain and itch are bothersome for children with EB, and can greatly affect their quality of life. Many factors contribute to the experience of pain, including what we think, how we behave and how we feel about pain (see diagram below). Children may experience physical and psychological effects, and families may experience emotional and social consequences as a result of pain and associated disability. It is important to know how to assess, prevent and treat your child's pain, often right from birth.



Assessing Pain

An accurate assessment of pain is essential to determine the best pain treatment. It is important to know where the pain is, what it feels like, how often it comes (or if is it there all of the time), and how strong it is. Pain assessment can also help to distinguish between pain and related feelings of discomfort, fear, and anxiety.

Because pain is individual to each child, assessment is not easy. Like sadness or fear, only the individual who is experiencing it can tell how intense it is or how much it bothers them. Only your child truly knows how the pain feels. Unlike reading a temperature with a thermometer, there is no objective measure to tell us how much pain someone has. However, we can use a variety of methods in combination to get a reasonably accurate assessment of pain.

How Do I know My Child is in Pain?

There are three ways to measure pain intensity: self-report, observer report, and physical changes. None of these is perfect. However, these methods can give parents and caregivers a good idea of the amount and type of pain a child has.

1. **Self report** – what your child says about their pain
   This method can only be used if your child can respond verbally or if they are able to understand the concept and can point (eg. to a written numerical rating scale or facial images). It involves asking your child directly about pain. These questions include:
   - Do you have a hurt/owie/pain?
   - Can you show me where it hurts? Does the hurt go anywhere else on or in your body?
   - When did the hurt start? How long has it been there?
   - Do you know what might have started it?
   - How much does it hurt? (Example 1, Numeric Rating Scale - on a scale of 0 to 10, 0 meaning no pain and 10 meaning severe pain, how strong is your pain? Example 2, Faces Pain Scale Revised (attached)
   - What helps to take away the hurt? (Medicines you've had before, heat/cold, playing with your friends?)

2. **Observer report** – e.g. a parent or caregiver
   Changes in your child's behaviour may indicate pain. There may be changes in facial expression, body movements, and the way your child cries. There are behavioural pain scales to help measure pain intensity more systematically to help monitor how severe the pain is, and whether treatments are effective e.g. the FLACC® scale or FLACC-revised for non-verbal children (see below).

Behaviour	0	1	2
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant quivering chin, clenched jow
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting, back and forth, tense	Arched, rigid or jerking
Cry	No cry (awake or asleep)	Moans or whimpers; occasional complaint	Crying steadily, screams, sobs, frequent complaints
Consolability	Content, relaxed	Reassured by touching, hugging or being talked to, distractible	Difficult to console or comfort

3. Physical changes – how the body reacts

Pain can be assessed by measuring changes to the body's physiological responses. An increase in heart rate, blood pressure, breathing rate and presence of sweating, can be caused by pain. However, measuring pain using physical changes alone is not reliable, and should be reserved for when other methods of assessing pain are not possible.

Factors influencing pain assessment

- The emotional context within which your child experiences pain is extremely important. Some children may be depressed, confused, worried, frustrated or angry about their pain. These emotional factors can have an impact on the amount and intensity of pain.
- Children may not want to talk about their pain. Children are sometimes taught that they should endure pain and that crying is a sign of weakness. These beliefs, which can apply to all cultures and ages and to all genders, may affect the way your child expresses and reports pain.
- A child may play down the extent of pain to please her parent or other caregivers. Children may also play down the extent of their pain out of fear. Your child may think that if she reports her pain as severe, she will have to stay longer in the hospital, away from her family, friends, and home.
- The way children are asked about pain may direct them to an answer. For example, asking "that doesn't really hurt, does it?" may cause your child to under-report pain. If your child is not asked about pain, she may say nothing even though she has pain. Conversely, frequently asking about pain can increase child's focus and attention on pain. We often suggest families have a check-in time e.g. after school or in the evening, where time is set aside to discuss.
- Before assessing your child's pain, let them know that it is important to be honest about their pain so that we can help. Children should feel they are part of the process of pain assessment and relief. Believing what your child reports helps give them and a sense of control that is ultimately beneficial.

Preventing and Treating Pain

Once you have assessed your child's pain, you can begin to plan for how to prevent and treat it. The plan may include psychological strategies, physical therapies, and/ or the use of medicines (pharmacology), which can all help relieve pain in children. Regularly assessing your child's pain will help you monitor how effectively the pain is managed.

Psychological strategies

Because emotion affects pain, the way a parent speaks can have a great influence on how their child responds to pain. The language that parents use should be as hopeful as possible and should not contain anything that your child might consider judgmental or doubtful. Acknowledging your child's pain, and being honest about what is happening, may help to reduce the distress and intensity of your child's pain. Children can be taught relaxation techniques to help them cope with pain. Breathing, imagery, visualization and distraction, are all helpful pain-coping strategies. Other techniques such as mindfulness meditation and cognitive behavioral therapy have been shown in research to greatly help children with chronic pain. Once learned, these strategies can be very effective in reducing pain

and other life stressors. Many hospitals have dedicated staff, often called Child Life specialists and health psychologists, who can help you and your child with developmentally appropriate strategies and resources.

Despite best efforts in managing pain, a pain-free existence may not be possible. When this happens, the focus turns to helping your child cope so that they may better manage normal activities of daily living such as attending school, sleeping well, participating in age-appropriate activities, even when they have pain.

Physical strategies

Physical therapies typically consist of methods such as heat, cold, touch and exercise but must be tailored to individual needs of child with EB.

As you can see, there are many strategies to help relieve pain. Finding the right combination can take time. Making adjustments for maximum pain relief requires re-assessment and a certain amount of trial and error in terms of finding strategies that work best for your child. Health care professionals will continue to work with you and your child to help relieve pain and improve quality of life.

Medication

Whether or not you use medication will depend on how severe your child's pain is. Often a combination of medication is required. Acetaminophen (e.g. Tylenol®) or Non-steroidal Anti-inflammatory Drugs (NSAIDs; e.g. ibuprofen) or both might be helpful for mild to moderate pain. For more severe pain, stronger pain medicines like opioids (e.g. morphine) might also be required. The decision to use one particular type of medicine over another will depend on the age of your child and the severity and type of pain. This should be planned together with your healthcare provider. Some of these medicines require a prescription from the doctor. For most pain, doctors and nurses use a step-up approach to target the type of pain and how bad it is.

Prevention is better than treatment. It is important to give pain medicines regularly to get on top of the pain and to prevent it from becoming unmanageable. The right dose of pain medicine for a child is determined by your doctor or nurse practitioner according to your child's weight with the goal of reducing the pain with the fewest side effects.

Special Considerations for Children with EB

Baths and Dressing Changes

Bathing and dressing changes are often painful, anxiety provoking experiences for children with EB. Pain medications and psychological strategies (e.g. Distraction, visualization, breathing exercises) should be used to prevent and minimize pain and fear. Pain medications may be taken by mouth (oral) or applied to the skin (topical) and may be given 20-30 minutes before the wound care routine. It is important to involve children in their care as much as possible. For example, allow them to help prepare dressing change materials, and to control for environmental factors that may be affecting pain (e.g. Reduce drafts, keep room warm). For older children cognitive behavioral techniques are strongly encouraged. Speak to your health care team about finding someone trained in these techniques, such as a psychologist or social worker.

Pain in the Gastrointestinal Tract

The gastrointestinal tract of children with EB is often a source of discomfort. Ulcerative lesions, acid reflux (gastroesophageal reflux disease), chronic constipation and esophageal strictures are the most common causes. Prevention is very important: a gastroenterologist and dietitian should be consulted to direct therapy (e.g. Optimal nutrition, use of stool softeners). Topical pain medication such as analgesic mouth rinses have shown to be helpful for mouth pain. Oral pain medications and psychological strategies are also important strategies to consider.

Joint and Bone Pain

An assessment and treatment plan developed with you, your child and health care team, including a physiotherapist and occupational therapist, is recommended. Staying active helps maintain bone health and reduce pain.

Eye Pain

Eye pain in children with EB is usually caused by corneal abrasions. Comfort measures such as avoiding bright light, lubricating eye drops and mild pain medications such as Acetaminophen and Ibuprofen, should be considered as needed.

Special Consideration for Pain in Infants and Very Young Children

For children with severe forms of EB, pain often starts immediately. Health care providers will use pain measurement scales specially developed for infants. A prescribed sugar solution called Sucrose can be used alone or together with other pain medications and other comfort measures such as swaddling, music, gentle rocking, may be used to prevent and reduce pain from procedures (such as dressing changes and bathing).

Pruritus (Itch)

Itch is often the most bothersome symptom for children with EB. There are several preventive measures that can be taken such as: drinking enough fluids and good skin care to prevent dry skin, good nutrition to promote healing, avoiding irritants (e.g. rough clothing) and scratching (short nails, covered skin) and avoiding overheating. Cognitive behavioral therapy can also be helpful to reduce scratching behaviors that may become a habit. Some pain medications, such as opioids, can also cause itch so it is important to balance pain relief with possible side effects.

Post-Operative Pain

Children with EB may need to have operations related to their diagnosis or for an unrelated reason. It is important that a pain management plan (for before, during and after the operation) is discussed and developed ahead of time with your healthcare team.

Dental Care in EB

Peter Judd, BSc, DDS, D Paed, MSc, FRCD(C)

Oral health is an important part of general health. Therefore, an unhealthy mouth can lead to greater health concerns. Avoiding disease through prevention is the most effective way to deal with tooth decay and infections or diseases of the mouth. Part of an effective prevention program is to work closely with a dental team dedicated to seeing each child have the healthiest mouth possible. The first important step is knowing at what age a child should first see the dentist.

When should your child first see the dentist?

A child should visit the dentist by six months of age or not later than when his/her first tooth erupts. Why?

- Seeing the child this early allows the dental team to provide to the parent/caregiver valuable information that can keep a child's mouth healthy even if there are no teeth in the mouth yet.
- What happens with this first appointment?
- The parent/caregiver is given guidance about what foods/drinks can harm the teeth and how to promote a healthy mouth.
- The parent/caregiver is shown the correct way to brush the teeth, when to brush and types of toothbrushes to use.
- The importance of regular dental visits is discussed and the next dental visit is arranged.

When is the second appointment (recall appointment) scheduled and how often should a child be seen?

This all depends on the child's risk for dental disease. A child judged to be at high risk for developing tooth decay should be seen every 3-4 months. Those who are not judged to be high risk should be seen every 6 months.

What happens at the recall appointments?

- Teeth are cleaned, if necessary
- Fluoride varnish is applied to help prevent decay
- Tooth cleaning techniques are reinforced and information to promote good oral health is provided

Typical treatment needs for patients under 6 years old who only have their "baby teeth"

- The goal is to prevent dental disease
  - Professional cleanings
  - Fluoride varnish applications
  - Oral care instruction
- If tooth decay is present, the dentist will review the treatment options that are available

Typical treatment needs between age 6 through 12, when the adult teeth erupt and the baby teeth fall out

- Sealants, a protective coating that prevents tooth decay, may be applied to the biting surface of some back teeth
- Regular fluoride varnish applications
- Professional cleanings
- If tooth decay is present, the dentist will review the treatment options that are available

Typical treatment needs of the adult dentition, 12 years of age and older

- The prevention program continues for life
- Treatment of decayed teeth, as required
- Orthodontic care or extractions as required

When treatment of dental disease is required, there are options for how the treatment will be done. Options include general anesthesia, sedation or in the dental chair using freezing. The choice for how treatment is completed is determined by the severity and extent of the tooth decay, the age of the patient and the severity of oral bullae.

Itching in EB

Elena Pope, MD, MSc, FRCP(C)

Itch is a very common symptom in the EB population. The exact reason behind itching is unknown. However, persistent skin inflammation, skin overheating due to dressings, skin sensitization from the use of topical creams, and the use of pain medications such as morphine and codeine may contribute. The vigorous itch may lead to more skin blistering. Itch is often poorly controlled symptom in most patients with EB and has a significant impact on their quality of life.

Management/Prevention of the itching includes

Non-medical treatment options

- Change the topical routine (switching dressings or discontinuing topical antibiotics may be sufficient to bring the itching down to a manageable level)
- Avoid skin overheating (such as many layers of clothing)
- Use cooling compresses or blankets for temporary relief
- Moisturize after each bath or as often as possible to prevent dryness
- Apply on the skin only preparations that have a low risk for sensitization (e.g. no fragrances)
- Discuss with your doctor new skin preparations that you would like to use

Medical treatment options

Using medicines by mouth to treat itch is not very successful. There is a lot of trial and error as most patients have a variable response to treatment. Your physician may try various preparations and escalate treatment depending on the response.

Physical & Occupational Therapy in EB

Jamil Lati, HBSc, BScPT, MScPT and Lisa Lazzarotto, BA, BSc(OT)

Rehabilitation

Physiotherapists (PTs) and Occupational Therapists (OTs) work to optimize the health and function of each child as they experience interruptions to normal health. We also facilitate the child and family's ongoing rehabilitation needs through links to our community partners.

Hands

It is important that your child is given the opportunity to use their hands to explore their environment. This includes playing with a variety of toys and games, as well as playing with other children at a playground or in appropriate group activities.

Watch for blisters on the front of the hands (palm) as well as between fingers (web spaces). Blisters in these locations can result in scars that limit how much the fingers can straighten and how much each finger can move away from fingers next to it. If the doctor or therapist is worried about your child's fingers not being able to move properly, the Physiotherapist may give you some simple, gentle exercises to increase or maintain the movement your child has in their hand. Sometimes, plastic splints are made for your child's hand by the Occupational Therapist to help maintain movement.

To help to prevent loss of movement in your child's hands, you can wrap their hands with protective dressings, both around the hand and in between the fingers. The dressings can reduce the blisters as well as allow for healing of the skin while maintaining movement. Ask your child's nurse for a dressing that will not stick to any open wounds on the hand, especially if you are putting dressings in between the fingers.

Soft and comfortable protective gloves can be custom-made for your child's hands to help protect them from blisters and/or to cover the dressings they wear on their hands. The Occupational Therapist can help arrange for gloves to be made for your child.

Feet

It is important to encourage your child to walk despite ankle pain, foot pain or blisters that may limit the movement of their toes. Always choose comfortable, supportive footwear to reduce pain and skin breakdown.
Your child may benefit from seeing a Chiropodist (foot care specialist) who can determine if your child requires custom-made seamless shoes. Crocs (plastic slip on shoes), running shoes with wide toe boxes or shoes designed for diabetic feet will allow the skin to breathe while accommodating dressings and protecting the feet from trauma. Shoe size needs to be assessed regularly (e.g. every 2 months for a child between 1 to 3 years of age due to rapid growth during this period).

Sheepskin or gel inserts may reduce pressure on the foot.

Your child should wear socks that are seamless or turned inside-out with the seams on the outside to prevent rubbing on the skin.

General activity

It is very important for your child to keep up a good level of activity to maintain their fitness level. This will help your child to maintain strength, flexibility, and endurance. This will minimize some of the complications of EB such as muscular tightness, weakness and pain.

Simple daily activities include:

- Standing with hips and knees straight as possible
- Walking as far as they are able to
- Lying on their stomach if tolerated (for babies)
- Gentle stretching activities focused on reaching their hand around a water-bottle or doorknob
- Pushing their hands into the padded seat of their chair to promote wrist and elbow flexibility
- Straightening fingers, elbows and knees for two minutes to prevent contractures

Seating and Pacing

Although it is very important to maintain a level of activity and fitness, there are situations that require the use of mobility aids, including the use of a wheelchair. Social activities may be limited if a child is unable to keep up with their peer group or if the child does not have the endurance to participate. In these situations, pacing themselves, taking breaks, and using a mobility aid will maximize your child's opportunities. However, it remains vital that your child continue to maintain a comfortable level of activity as recommended above.

School

Most children with EB attend regular schools but may require special accommodations. The principal and teacher will need as much information about EB as possible to help your child to be successful. You should be prepared to discuss some of the following topics with the school:

- In school Occupational Therapy assessment
- Writing accommodations
- Seating
- Pacing
- Gym and recess

Please refer to Epidermolysis Bullosa, A Guide for Parents & Schools available from DebRa of America, Inc. at www.debra.org for information regarding education, adaptations and suggestions.

Clothing

Your child may be more comfortable wearing thin, soft, loose fitting clothing over their dressings. Seams next to skin may cause friction and blistering. To prevent this, turn the clothes inside out so the seams are on the outside.

Your child should avoid clothing that has tight elastic bands, sharp buttons, snaps and zippers. Remove clothing tags and labels before use. Wearing soft undergarments or bandages may be necessary to protect the skin. Clothing should be easily to put on and taken off and not be tight or rub/shear at the head, ears and armpit areas.

For infants, place cotton mittens over their lubricated hands and feet to reduce them from scratching or rubbing their face. To protect elbows from rubbing, cut the toes off a large pair of socks and then pull the socks over the elbows. If your child gets blisters from the sides of the crib, pad their elbows, heels and knees with gauze sponges secured by rolled gauze. Full sleepers, which include feet, are recommended. For young children who are learning to crawl and walk, consider adding foam padding to the clothing in the knee and elbow area.

Cardiomyopathy in EB

Irene Lara-Corrales, MD, MSc

Cardiomyopathy is the name given to a disease of the heart muscle affecting the way the heart functions. There are different types of cardiomyopathy. Patients with EB, particularly those with junctional and dystrophic types of EB, are at a higher risk of developing a specific type of cardiomyopathy called **dilated cardiomyopathy**.

In dilated cardiomyopathy, the heart chambers become enlarged or dilated. The heart muscle becomes weak, making the pumping of blood to the rest of the body very difficult. Mild cases of cardiomyopathy might not be detected because they may not present any symptoms, but severe cases do present with symptoms, and in severe cases may even lead to death. As dilated cardiomyopathy may develop without any symptoms, it is important to have regular heart checks, particularly in junctional and dystrophic EB where this condition has been reported.

What causes dilated cardiomyopathy in EB?

No clear cause of dilated cardiomyopathy has been identified in patients with EB. There have been several possible causes implicated such as low mineral/nutrients levels, viral illnesses, chronic anemia and some medications, but no single clear cause explains all cases. The possibility of a genetic predisposition has also been proposed, but this has not been identified yet.

What are symptoms that can be associated with dilated cardiomyopathy?

The most common symptoms are:

- Shortness of breath when doing some effort
- Shortness of breath when lying down or at night waking up the patient
- A decrease in the ability to be active, walk or exercise
- Swelling of lower legs, ankles and feet
- Fatigue
- Weight gain, cough and congestion due to fluid retention
- Feeling your heart racing or slowing down
- Dizziness or feeling lightheaded

How is the diagnosis made?

Tests such as an electrocardiogram (examines the electrical activity of the heart) and an echocardiogram (an ultrasound of the heart) are the two heart tests that can identify patients with dilated cardiomyopathy. In EB patients, the medical history and physical exam might not identify early symptoms. Bloodwork (hemoglobin, albumin, free and total carnitine, selenium, zinc) are also part of the work-up for patients suspected of having dilated cardiomyopathy.

Treatment

Sometimes medications are useful to improve the function of the heart, especially if the problem is identified early.

Prognosis

Early diagnosis and management can change the long term outcomes of dilated cardiomyopathy. Monitoring of symptoms, and getting regular heart ultrasounds by cardiology helps us detect changes early so we can intervene before symptoms get worse.

Renal Disease in EB

Elena Pope, MD, MSc, FRCP(C)

Renal impairment may occur in some of the severe subtypes of EB. In general, renal problems can be due to urinary tract involvement or direct kidney damage.

Renal Disease due to Urinary Tract Involvement

Urinary tract problems occur in approximately 12% of cases of cases of severe Junctional EB and approximately 8% of Recessive Dystrophic EB. Blistering or ulcers may occur anywhere along the urinary tract causing multiple problems:

- Scarring and ultimately permanent narrowing of the urinary passages
- Chronic obstruction of the urinary flow leading to backing up of the urine, stretching the ureter (a tube connecting the kidneys to the bladder); chronic dilatation of the ureter (**hydroureter**) causes dilatation of the kidneys (**hydronephrosis**) and ultimately kidney failure
- Recurrent urinary tract infections that may exacerbate the chances of developing hydroureter

Renal Disease due to Kidney Damage

Repeated kidney damage either due to hydroureter and/or chronic systemic infections (post-infectious glomerulonephritis, Ig A nephropathy) will result in permanent renal scarring. Renal scarring can, in turn, severely affect the function of the kidney. It is estimated that ~12% of patients with RDEB will have renal failure by age of 35 years.

It is important for at-risk patients to be assessed annually by performing a urine analysis and kidney function. For older patients, keeping the blood pressure in the low range and potentially using medications that will have a protective role for the kidneys and may be beneficial. Circumcision may be medically indicated in some cases to reduce the chances of strictures due to blistering.

Haematological (Blood) Issues in EB

Melanie Kirby, MD

What is anaemia?

Anaemia is the most common blood issue in EB. **Anaemia** is when not enough red blood cells are made. Red Blood cells carry oxygen to the tissues and organs. Oxygen is required for all body functions including growth and wound healing. Anaemia is detected as a low haemoglobin level on a blood test.

What are the signs and symptoms of anaemia?

A person who is anaemic has low energy. The heart rate increases to try to compensate for the lower numbers of red blood cells in order to deliver oxygen to the organs and tissues. One can feel dizzy or faint when standing up and may experience shortness of breath. Headache is also a frequent complaint. Long standing anaemia will affect growth and development and impair healing of skin wounds.

Why do persons with EB have anaemia?

Individuals with EB develop anaemia because they lack many of the building blocks required to make red blood cells, including iron, proteins, vitamins, and trace metals. Normally, these nutrients are absorbed from food through the gut lining. In EB, the gut lining is inflamed and absorption of iron, proteins, vitamins, and trace metals is reduced. Sometimes persons with EB also have overall poor nutritional state and low protein intake. As such, the proteins that help with absorption are very low and so iron is not transported properly. Proteins are also lost in the fluids that are lost from skin wounds.

What can be done to reduce the degree of anaemia?

All measures to improve nutrition should be taken. These include supplementation with iron, folic acid, Vitamins E, D, K and B12. Nutritional requirements will be covered elsewhere.

Iron Supplementation
Oral iron supplements should be attempted daily using a preparation that is best tolerated by your/your child. They may be taken with food, except for cereals and dairy products since these foods decrease the absorption of iron from the gut. Vitamin C or fruit juices enhance absorption of iron. Iron supplements may cause upset stomach and constipation. As such, even if oral supplements can be tolerated, the iron absorption may not be adequate to treat the anemia.

If oral iron supplements are not tolerated or do not result in an increase in haemoglobin, then intravenous (IV) iron may be given. There are a few different preparations available and your doctor will work with you to decide which one is safer. An occasional complication of IV iron is an allergic reaction; as such IV iron is given in the hospital, often in an outpatient setting, with health care team in attendance. IV iron goes straight into the bloodstream so it does not require absorption as it bypasses the gut. The frequency of iron administration will be determined based on the response and haemoglobin level.

Erythropoietin, a protein normally produced by the kidney, may be added to IV iron. Normally, erythropoietin is produced when haemoglobin levels are low. If erythropoietin levels are inappropriately low in you/your child's blood, adding it to a regimen along with IV iron may be necessary. Erythropoeitin is given as an intramuscular injection 1-3 times a week or once a week depending on the preparation.

Note: if your child severely symptomatic due to a very low hemoglobin level (for example, fainting, low heart function), a blood transfusion may be required. Your attending physician will work with you to determine the need.

Monitoring for Anaemia

We perform the following to monitor for anaemia:

- **Complete Blood Count (CBC)**: Haemoglobin level and small size of red cells are clues to iron deficiency
- **Ferritin**: usually low in iron deficiency. In EB, ferritin is not reliable because of ongoing inflammation which can increase ferritin levels
- **Serum iron levels**: reliable
- **Transferrin**: is the binding protein of iron. Not reliable for assessing for iron deficiency due to low protein state of patients with EB

Other Blood Issues in EB

Both bleeding and clotting may be an issue in patients with EB. Normally, clotting proteins and anti-clotting proteins are present in balance in our blood. When proteins are lost from the damaged gut lining and skin of patients with EB, this balance can be affected. Symptoms of bleeding or clotting will depend on which of these proteins are present in lower amounts in the blood stream. At the time of presentation, blood tests will help to determine what the underlying cause is, and treatment will be started. Treatment may involve correcting the imbalance using a blood product called Fresh Plasma or other clotting proteins derived from blood donor pool.

Skin Cancer in EB

Elena Pope, MD, MSc, FRCP(C)

Certain subtypes of EB are at higher risk of developing a type of skin cancer called **squamous cell carcinoma (SCC)**. In contrast with skin cancer in the general population, skin cancer in EB tends to affect younger patients starting in their second decade of life, occurs in more than one site at the same time (multifocal) and has a more aggressive course.

Who is at risk?

Although most of the severe EB types can present with skin cancer, it more commonly affects patients with generalized RDEB. Other subtypes that can be affected are DDEB and non-Herlitz JEB and rarely, Dowling Meara type of EBS. For all other subtypes there is no increased risk of skin cancer above what is expected in the general population.

How does skin cancer present in EB patients?

Skin cancer usually presents with a chronic, non-healing ulcer. As ulcers and chronic wounds are common in severe EB subtypes, a high degree of vigilance is needed for both patients/families and their health care professionals. Signs that should increase suspicion for skin cancer are:

- Increase in the wound healing time typical for patient
- Wound feels or looks different from other wounds
- Wound size increases rapidly
- Wound is more painful than usual
- There is persistent, hard, yellow-gray tissue next to a chronic wound

How do you prevent skin cancer or discover it early?

- Ensure regular (every 3-6 months) full skin inspection in high risk individuals by a specialized team
- Seek medical attention as soon as possible if any of the suspicious changes are noted

How do you treat skin cancer in EB patients?

Patients need to be looked after in centers specialized with skin cancers in patients with EB. Treatment requires surgery to remove the cancerous area as completely as possible. In addition, patients will require more investigations to detect the extent of the cancer (spread to lymph nodes, lungs, etc.). Occasionally, the tumor may be so extensive that more invasive procedures may be required (such as limb amputation).

What is the prognosis of the patient diagnosed with cancer?

Prognosis is guarded in patients diagnosed with skin cancers. If the cancer is limited to small areas, complete excision and close follow-up every 3 months is recommended. Patients with large cancers, multiple affected sites, lymph node involvement and/or metastasis have a very poor prognosis.

Bone Health in EB

Elena Pope, MD, MSc, FRCP(C)

Patients with EB are at higher risk for “weaker” bones. This risk is proportionate to the disease severity. Potential explanations for this complication are:

- Low intake of calcium rich foods
- Low vitamin D levels due to lack of exposure to the sun (bandages and/or not spending time outside)
- Lack of mobility

The bony problems in EB may range from:

- Osteoporosis (decreased bone density)
- Fractures (either occurring spontaneously or after trauma)
- Bony pain without radiological evidence of fractures

The risk of bone weakness increases dramatically after patients become wheelchair bound or after episodes of prolonged immobility (for example, admission to hospital).

Bone density has to be monitored on a yearly basis by performing a special scan called DEXA scan.

Measures that prevent bone loss are:

- Maintain a daily routine of walking
- Take supplements such as Vitamin D and calcium
- Maintain a good diet

Hair & Nail Involvement in EB

Lauren Briggs, BSc

Some subtypes of EB are associated with hair and nail abnormalities.

Nail involvement

The severity of the findings varies from person to person but is dependent on the type of EB they have. In some cases, it may be mild; however, the nail involvement caused by EB may be disabling in others. Blistering in the nail bed due to minimal trauma (cutting of the nails) can cause nails to appear ridged, grow misshaped or fall out. Chronic open sores around the nails may lead to infections.
Early nail involvement may be a sign of a more severe or progressive form of EB.

Hair involvement

EB can also cause hair changes. EB causes blistering of the scalp, leading to inflammation and scarring around the hair follicles leading to permanent hair loss in some areas. In addition, EB can cause the hair to become more prone to damage to common hair practices.

Eye Problems in EB

Asim Ali, MD, FRCS(C), Ali El Hamouly, BMSc and Hamza Sami, HBMSc

In epidermolysis bullosa (EB), the eye can often be involved and therefore, regular assessment by an ophthalmologist may be required. Below is a list of common eye issues related to EB:

Common symptoms

- Red, watery eyes
- Eye pain
- Photophobia: sensitivity to light

Eye issues

The **eyelid** protects your eye from physical debris and lubricates it to keep it moist. In EB, patients can get:

- **Eyelid blisters**
- **Ectropion**: the outward turning of the eyelid e.g. due to scarring of the skin in EB.
- **Entropion**: the inward turning of the eyelid due to scarring inside the lid. Both entropion and ectropion may require surgery to correct.

The **conjunctiva** is a thin membrane that cover the inside of your eyelids and the sclera. It is involved in eye lubrication and protection of the eye from infection. In EB, the conjunctiva can be affected by the following:

- **Conjunctival blisters**
- **Conjunctival edema**: swelling of the conjunctiva due to fluid build-up from inflammation and irritation
- **Conjunctival injection**: reddening of the conjunctiva due to irritation
- **Symblepharon**: the attachment (due to scarring) of the conjunctiva on the inside of the eyelid with that over the outer, white portion of the eye, the sclera. This may require surgery to remove the scar tissue and amniotic membrane grafts to promote normal healing.

The **cornea** is the clear curved layer of the eye that lies above the coloured part of your eye, the iris. It part of your vision system that allows light to be focused on the back of your eye. In EB, the following can occur to the cornea:

- **Corneal abrasion**: scratching of the cornea, leading to marked pain, blurry vision & tearing. This will require antibiotic eye drops to prevent infection, and the use of a patch or a bandage contact lens to reduce pain and speed healing. Sometimes dilating drops (cycloplegics) are given to reduce pain.
- **Corneal pannus**: formation of scar tissue on the cornea that can block vision and cause irritation. In severe cases this can be removed surgically.

The Psychosocial Impact of EB

Nimrita Aujla, MSW, RSW

Initial Diagnosis

Learning that your newborn has been diagnosed with EB can be an overwhelming experience. It is important to know:

- Everyone reacts differently and may experience a range of emotions including guilt, sadness, and anger. These feelings are common.
- Caregivers may experience the stages of grief and loss which include denial, anger, bargaining, depression, acceptance (Kubler-Ross Model).
- Mothers should be assessed for post-partum depression. Speak to you family physician about your mood.
- There will be many unknowns which can create feelings of fear. Some of your questions or concerns may not be answered right away as the practitioners may not have all the answers.
- Educate yourself about EB by asking questions, joining a support network (DEBRA Canada) and reading medical literature. Be cautious of information found through the internet. Do not give anyone money who says that they can cure EB. Talk to your medical team before trying any new products.
- You may have lots of medical appointments at the beginning. Write down questions that you have for the medical team. Keeping medical information organized will help you during these appointments. Bring a support person with you who can help you ask follow-up questions and remember discussions after you leave.
- Build a support network for your family. This network can consist of close family and friends and the medical team.
- Take care of yourself so you can take care of your newborn. As they say in airplanes, put your oxygen mask on before trying to help someone else.

Newborn Stage

- Attachment is crucial between a caregiver and newborn, not only for your newborn to feel safe but also on your newborn's future relationships. An important factor of attachment is positive physical contact. This can be achieved by holding, hugging, kissing and face-to-face contact.
- Don't be afraid to hold and touch your newborn. Modifications will need to be made including not picking up the baby by the underarms and holding the baby by using a soft blanket underneath them. Also pat rather than rub the baby.
- You will need to learn how to care for your newborn and become the expert in what works and what does not (i.e.: the best dressings). Figuring out what works and what does not will be a trial and error process.
- Do not be afraid to ask your medical team questions.

Pre-School Stage

- During this stage, children are exploring their environments and learning how to sit and eventually crawl around on their hands and knees.
- During these times of exploring, children will develop blisters in the areas where friction occurs (i.e.: hands and feet). Talk to your medical team about bandaging techniques for these areas to minimize the friction.
- Understandably, you want to protect your child from injury and pain. However, blisters cannot be completely prevented so it is important to allow your child to explore their environment in order for them to achieve their developmental and social milestones.

School Age

- Caregivers often have lots of fears around their child entering the school system. Common questions that come up include; “Will my child be bullied?”, “What if my child can’t participate in physical education?”, “What modifications can be made for my child in the classroom?”
- Create a partnership with your child’s school. Prior to your child starting, meet with staff to discuss what EB is and what your child’s needs are. Each school is different and provides different accommodations. Discuss your concerns and work together to put plans into place should certain situations occur including emergency medical situations. Some schools may ask for medical documentation.
- Children with EB may have increased school absences due to illness and medical appointments. Should this be the case, work with your child’s school to develop a plan so they don’t fall behind.
- EB is not associated with learning disabilities.
- This is a good time to start talking to your child about what EB is. Please see below, “Talking to Your Child About EB”.
- Have a discussion with your child about what to say if someone asks him/her about their skin. Preparing your child for these situations can help them be confident when responding to others questions.
- Enhance your child’s self-esteem (let them develop their independence, allow them to develop competence, allow them to develop connections with others).

Adolescence and Beyond

- This is a very difficult time for anyone especially as image and relationships become a primary focus.
- Physical limitations may prevent adolescents from participating in activities that their peers may be participating in.
- Help and encourage your adolescent to discover a hobby. This can increase their self-esteem and confidence as well as their social network.
- It is important for your adolescent to be as independent as possible. This helps them achieve a sense of control over their own lives. This can be very difficult for caregivers, who have for years been responsible for many aspects of their child’s care.
- Even if it is not possible for the adolescent to take over their medical treatment, it is important for the treatment to be guided by the adolescent and to ensure that they are aware of their medical treatment/medical appointments/procedures/medication etc. Caregivers should discuss these topics with their adolescent openly.

- It is also important for everyone to be aware that:
  - Regular skin checks by the medical team are imperative
  - Squamous cell carcinoma is a serious concern

*** It can be frustrating to have multiple appointments where it seems like nothing new is happening. However, regular medical monitoring is very important as physicians are trained to see things that are concerning that we may not notice. There are also things going on in our body that we cannot see which need to be monitored through blood work and other tests. This is important as some things can quickly spread to more serious complications.

Siblings

- Siblings may feel that they are not receiving the same amount of attention compared to their affected siblings. Spend one on one time with siblings.
- Siblings may feel a sense of anger or jealousy towards their affected sibling. It is important for them to know that this is a common feeling however, it is important for them to be aware of EB and how it affects their sibling.
- It is important to speak openly and honestly with siblings about EB as not knowing may lead them to imagine the worst and worry about their siblings.

Talking to Your Child About EB

- Speaking to your child about EB is important. You may want to protect your child however, they can
- often sense when something is wrong. The unknown can cause significant fear and stress and lead to imagining the worst.
- Discussions should be based on the child's age.
- Prepare them for hospital visits and medical procedures by being honest about what will happen. It is ok for them to know that a test may hurt.
- Use simple ways to explain EB (i.e.: delicate skin).
- Let them know EB is not contagious.
- Children will commonly ask why me? You can respond by telling them "no one knows why but we know it is no one's fault". They may ask, "Will I get better?" You can respond by telling them, "EB doesn't go away, that is why we go to see the doctor and do things at home to try to make the EB as manageable as possible".
- As they get older, it is important to start talking to them about:
  - The EB subtype they have been diagnosed with
  - The different subtypes of EB
  - Why they are seeing certain doctors
  - Why they are they taking certain medications
- Have them be part of the medical appointment and encourage them to speak up and talk to the doctor about how they are feeling.

Monitoring for Complications in EB

Elena Pope, MD, MSc, FRCP(C) and Irene Lara-Corrales, MD, MSc

Test/ Intervention	EBS	JEB	DEB
Skin swabs for culture and sensitivity (if infection is suspected a skin biopsy is more reliable than a skin swab as most patients are chronically colonized with multiple bacteria)	As needed	As needed	As needed
Blood Investigations ■ **Hematology:** CBC, iron studies ■ **Chemistry:** calcium, phosphate, zinc, selenium, electrolytes, renal function tests, carnitine, PTH, Vit D	Once per year (Annually)	Once every 6 months (two times per year)	Once per year (Annually)
Urine testing Urine dipstick for protein and blood	Once per year (Annually)	Every 3 months or more frequently if needed	Every 3 months or more frequently if needed
Imaging ■ DEXA scan ■ Renal Ultrasound ■ Echocardiogram ■ Bone x-rays	As needed	Once per year (Annually)	Once per year (Annually)
Nutritional Assessment	Once per year (Annually)	Every 3 months	Once per year (Annually)

Footwear Resources for EB Patients

Michelle Lee, RN, BScN

Footwear

To prevent foot diseases, it is important that your child practices good foot hygiene. Socks should be changed daily, and ideally, shoes should be alternated daily so that they can be aired out. Your child's feet should be washed and dried thoroughly every day, including the areas between toes. Let your child's feet air out; do not keep them in shoes or bandaged all day. Cut your child's toenails short. You may choose to apply creams and powders to keep the skin soft and dry, as well as to prevent infection and odor.

To prevent blistering, cut the elastics on socks and have your child wear thick cotton socks, ideally two pairs, to reduce friction. Dress your child in soft, loose-fitting shoes with rounded toes and lots of extra room for padding. Take note of the location of the seams, and use padding to prevent blistering in these areas. The soles should be flat and flexible, with some form of heel support. To prevent movement or slipping of the foot inside the shoe, laces or straps may be recommended. Shoes should not be made of plastic or other synthetic materials; permeable leather soles are often a good choice, as this material will help keep the foot cool and dry. Many children with EB choose to wear sheep-skin lined slippers, ballet shoes or moccasins instead of conventional shoes.

If your child's feet are severely affected by blisters, he or she may be more inclined to walk around without shoes, and just have extra padding around their feet.

Test/ Intervention	Product Name	Description	Provider
Foot care	Blister Shield	Powder to be used in socks to reduce rubbing	TwoToms LLC www.2toms.com
	Burrows Solution/ Drysol/ Arrid XX dry antiperspirant	Aluminum-containing products recommended by EB families	
	Domeboro Soaks	Aluminum product reported to dry up blisters and quicken healing	www.drugs.com/pdr/domeboro_tablets.html
Shoes	Bilby Shoes	Custom shoes with soft leather tops and sponge lining	Bilby www.bilbyshoes.com *On the site, go to "Products" and "Medical Grade and Extra Depth" for special EB shoes*

Test/ Intervention	Product Name	Description	Provider
Shoes	Moccasins	Soft and can accommodate dressings	Minnetonka Moccasins www.pueblosouthwest.com/Moccasins.html
	Ugg boots	Sheepskin lined boots, shoes and slippers	UGG Australia www.uggaustralia.com Available at many retail outlets
	Elefanten	Soft, flexible shoes	www.elefanten.de/Elefanten/en/Elefanten_index.html
	Berber Fleece Slippers	Fleece slippers (Item # TA48887) Ankle cuff slippers (Item # TA48940)	LL Bean www.llbean.com
	Pedors	Adjustable closure, soft, flexible to accommodate dressings. May need additional soft insole for padding.	Pedors www.pedors.com
	Pediped	Soft and flexible, easy to put on	Pediped www.pediped.com (800) 880-1245
	Robeez	Soft, flexible shoes	Robeez www.robeez.com
	Preschoolians	Soft, flexible shoes	Preschoolians www.preschoolians.com
	Saltwater Sandals	Soft, flexible shoes	Hoy Shoe Company www.saltwater-sandals.com 4970 Kemper Ave St Louis, MO 6313 (314) 772-0900
	Adidas Climacool	Trainers with mesh lining and shock absorption, which allow air circulation	Adidas www.adidas.com/home/ca
Socks	Swedish Moccasins	Leather soled socks (Item # ZA18611)	Hanna Andersson www.hannaanderson.com
	Silver Sock	Silver-containing socks which prevent friction and regulate heat	Carnation Foot care www.silversock.co.uk

Many children with EB have successfully used shoes from the brands provided above; however, another option is to purchase custom footwear. Phillip Watson is an orthotist with experience crafting shoes for EB patients, and works for Ambulatory Footwear in Dundas, ON (www.ambulatoryfootwear.com).