

Reframing the Care of Children With Epidermolysis Bullosa Through the Lens of Medical Trauma

Rebecca Cross Bodán

ABSTRACT: Epidermolysis bullosa (EB) is a group of rare inherited genetic connective tissue disorders causing blisters and skin erosions, in addition to a number of other extracutaneous complications. There is currently no Food and Drug Administration-approved treatment for EB, and the current mainstay of disease management involves frequent, prolonged, and extensive wound care, which is often traumatic for the child and caregivers alike. Because of the potential for extensive wounds and other complications, children with moderate to severe forms of EB experience significant amounts of medical trauma related to their condition, both in a medical setting and in the community. Exposure to pediatric medical trauma has been linked to poor health outcomes and increased psychosocial sequelae. Thus, this article seeks to provide an in-depth discussion of the potential sources of trauma children with EB face on a daily basis, along with some practical trauma-informed interventions that healthcare providers may consider implementing in an attempt to alleviate some of the suffering and build resilience among those affected by this potentially devastating and life-limiting condition.

Key words: Epidermolysis Bullosa, Pediatric Medical Trauma, Trauma-Informed Care, Rare Disease

Epidermolysis bullosa (EB) is a group of rare genetic connective tissue disorders. The primary feature all forms of EB have in common is skin blistering or erosions as the result of mild mechanical trauma (Dystrophic Epidermolysis Bullosa Research Association [DEBRA], 2018; Fine et al., 2014). EB is a lifelong condition, which typically presents at birth, with skin erosions and bullae (DEBRA, 2018). As a result of these wounds, and because there is no Food and Drug Administration (FDA)-approved treatment for the condition, those with EB require frequent, extensive, and prolonged wound care throughout their lives (Adni, Martin, & Mudge, 2012; DEBRA, 2018; El Hachem et al., 2014; Fine et al., 2014; Kirkorian, Weitz, Tlougan, & Morel, 2014; Mauritz et al., 2019; Pope et al., 2012; Schober-Flores, 2003). In most cases, parents or caregivers perform the complex wound care (Mauritz et al., 2019; Tabolli et al., 2010).

Whereas localized forms of EB tend to be limited to painful blistering on hands and feet, those with generalized forms can develop wounds on any part of the body, along with a number of systemic, extracutaneous effects (Fine et al., 2014). The systemic complications of EB can include corneal abrasions and dryness, esophageal blisters and strictures, oral blisters and ulcers, erosion of dental enamel, cardiomyopathy, constipation and rectal bleeding, anemia, fine and gross motor delays, mitten deformity, fusion of digits, contractures, and growth retardation, along with a number of other potential complications (Boeira et al., 2013; DEBRA, 2018; Feijoo et al., 2011; Fine, Johnson, Weiner, & Suchindran, 2008; Murat-Sušić, Husar, Skerlev, Marinović, & Babić, 2011). As a result of the many systemic complications

Rebecca Cross Bodán, FNP, PhD, Fullerton School of Nursing, California State University, Fullerton, CA.

R. C. B. wrote and edited the article.

Dr. Bodán is the mother of a school-age child born with severe generalized epidermolysis bullosa simplex.

The authors declare no conflict of interest.

Correspondence concerning this article should be addressed to Rebecca Cross Bodán, FNP, PhD, Fullerton School of Nursing, California State University, EC-642 800 N. State College Blvd., Fullerton, CA 92831. E-mail: rbodan@fullerton.edu

Copyright © 2020 by the Dermatology Nurses' Association.

DOI: 10.1097/JDN.0000000000000514

associated with generalized forms of this disorder, children with EB often require care from a large multidisciplinary team (Ahmad & Bruckner, 2014). They are also at a higher risk of developing psychosocial sequelae, such as depression or anxiety, along with impairments in quality of life compared with their healthy peers (Dures, Morris, Gleeson, & Rumsey, 2011; Margari et al., 2010; Pagliarello & Tabolli, 2010; Tabolli et al., 2010).

Perhaps, one of the lesser talked about aspects of life for children with EB is the frequent and repetitive exposure to pediatric medical trauma. The National Child Traumatic Stress Network (NCTSN) has designated the experience of trauma during childhood illness as an adverse childhood event (Pynoos et al., 2014). Children exposed to adverse childhood events are more likely to experience physical and emotional health problems as adults, compared with the general population (Felitti et al., 2019). A growing body of evidence shows that the exposure to chronic illness and medical traumatic stress in childhood can have both immediate and long-term negative impacts (Espeleta et al., 2019; Holbrook et al., 2005; Price, Kassam-Adams, Alderfer, Christofferson, & Kazak, 2016; Zatzick et al., 2008). The provision of trauma-informed care (TIC) can help mitigate this risk of traumatic stress by intentionally building resilience and informing families when intervention is needed early on, giving the child and family the greatest chance of being able to better cope with the stress and trauma of EB (NCTSN, 2014). Thus, the purpose of this article is to provide insight into the many potential avenues of medical trauma experienced by children with EB, while addressing how the multidisciplinary EB team can provide TIC by gaining an understanding of the forms of EB-related trauma and its impact on the child and family, while also promoting resilience and other protective factors for those affected.

PATHOPHYSIOLOGY AND EPIDEMIOLOGY OF EB

EB is thought to affect 1:20,000 live births, with approximately 500,000 affected people worldwide (DEBRA, 2018). The incidence and prevalence vary depending on EB subtype. There are four main types of inherited EB, which are differentiated based on the location of blister formation within the epidermis: EB simplex (EBS), which accounts for approximately 70% of all EB cases, with blisters mostly occurring at the basal layer; dystrophic EB (DEB), where the sublamina densa is affected; junctional EB (JEB), where the level of skin cleavage is the lamina lucida; and Kindler syndrome, where variations in the level of skin cleavage occur. Genetic forms of EB are inherited in either a dominant (EBS, dominant DEB) or recessive (rare types of EBS, recessive DEB, JEB, and Kindler syndrome) pattern (DEBRA, 2018; Fine et al., 2014). There are over 30 subtypes of the condition, with a wide range of phenotypes.

PEDIATRIC MEDICAL TRAUMATIC STRESS DEFINED

The NCTSN defines pediatric medical trauma as a “set of psychological and physiological responses of children and

their families to pain, injury, serious illness, medical procedures, and invasive or frightening medical experiences” (NCTSN, 2014). The child's reaction to medical trauma has less to do with the severity of the medical event and is more related to the child's perception of what is happening (Price et al., 2016). This serves as an important reminder to providers that relying only on the severity of an event or illness does not provide sufficient data to determine which children/families may be at a greater risk for developing psychological consequences as a result of medical trauma. There are, however, a number of known factors that may increase the risk of developing persistent traumatic stress reactions in children who experience medical trauma, including exposure to or experience of one or more of the following (NCTSN, 2014):

- Severe early traumatic stress reactions
- Experience more severe levels of pain
- Exposed to scary sights and sounds in the hospital
- Separated from parents or caregivers
- Previous traumatic experiences
- Prior behavioral or emotional problems
- Lack of positive peer support

A number of the above risk factors for persistent traumatic stress are inherent in EB. For example, for children born with moderate to severe EB, the traumatic events begin on the day of birth as they are cared for in the neonatal intensive care unit (NICU) with painful wounds (Bodan, 2016; Denyer, 2010). Pain is inherent to this condition, and pain management is often underutilized (Bodan, 2016; Goldschneider et al., 2014). When healthcare providers understand the potential causes and reactions to medical traumatic stress, they can assess, treat, and, in some cases, even prevent traumatic stress reactions during medical encounters, starting as early as the NICU.

PEDIATRIC MEDICAL TRAUMA IN EB

The reality for many living with EB is that much of life has the potential to cause trauma, whether that is trauma from wound care, medical procedures, or even harm from innocuous events in day-to-day life (see Table 1 and Figure 1); when one has EB, there is no escaping frequent, traumatic events. Although most providers who care for children with EB are well aware of the pain that these children face on a daily basis, the surgeries they often must undergo, and the need for extensive wound care (Table 2), a discussion of medical traumatic stress as it relates to EB has not been published to this author's knowledge.

Adapting the Pediatric Medical Trauma Model to EB

The current models for pediatric medical trauma are focused on relatively short-term medical encounters (Kazak et al., 2006; NCTSN, 2014). Children with EB have frequent encounters with the healthcare system; these models are applicable to these situations. What these current models do not address is in reference to the EB-related trauma that occurs in the home and community, as pain, injury, and trauma are a daily part of life for many with EB.

TABLE 1. Examples of Trauma Experienced by Children With EB

Types of EB Trauma	Examples
Wound care	Wound care occurs daily, or almost daily, and often takes several hours. The process (described more in Table 2) is extensive and often painful. Wound care is commonly performed by the parents or caregivers; thus, it is important to consider the impact of this trauma on the entire family.
Medical procedures	Children with moderate to severe EB often require medical procedures beyond wound care. Common procedures include esophageal dilations, oral surgery, G-tube placement, and hand surgeries. Although these can be traumatic for any child, they are inherently more risky for those with EB.
Medical providers	Minor medical tasks such as taking a blood pressure or venipuncture can be very damaging to the skin of a child with EB. Parents and patients must remain vigilant to decrease the likelihood of their occurrence; in short, going to clinic visits is often not a safe space for children with EB and can be very stressful for their caregivers.
School	Potential for school-related trauma will vary based on the age and abilities of the child. The simple act of sitting can be very painful. Eating is often slower and can result in episodes of choking. Typical playground activity is often not possible, but when attempted injuries can easily occur by being bumped into by another child, falling, and so forth, crowded school hallways are another source of concern. The risk of emotional trauma from bullying is high and can seriously impact a child's ability to perform well in school. Finally, the simple fact that many children cannot keep up with their peers and may feel left out or isolated can also feel traumatic for the child and family.
Community	Young children with EB can incur significant wounds from family or friends who simply hold them in the wrong way. Older children can experience trauma from their peers on playgrounds or when attending community classes or events. Many children will experience the emotional trauma of not being able to participate in events and activities that are deemed not "EB friendly." There are very few trauma free spaces for children with moderate to severe EB.

Sources: Ahmad & Bruckner, 2014; El Hachem et al., 2014; Fejoo et al., 2011; and Goldschneider et al., 2014. EB = epidermolysis bullosa.

TABLE 2. Wound Care Routine for Children With Moderate to Severe EB

EB Wound Care Steps	Description
Setup	Bathtub is cleaned and disinfected. Supplies are cut and set out on a designated space.
Bath	Often with additives such as salt, bleach, or vinegar (to limit "sting," decrease bacterial burden, and improve healing). Soaking in the bathtub is a necessary but often painful experience, depending on status of wounds and severity of disease.
Removal of bandages	Completed either before bath, in the bath, or after bath, depending on patient preference and wound severity. Wounds can often adhere to dressings, which adds to anxiety, anticipatory anxiety, and pain.
Transfer location	Children will either walk or be carried from the bath to the wound care space. For those who are unable to walk, this can put a significant physical strain on caregivers as the child grows.
Managing blisters and wounds	Draining and "flattening" of blisters with needles or scissors. Blisters left undrained will continue growing, which increases pain and infection risk. Removal of crusts and debridement of skin as needed.
Application of topicals and dressings	Application of topical oils, ointments, and/or creams, either directly to the skin and wounds or "buttered" onto specialized dressings (often silicone based). Dressings applied to active wounds and to areas at a high risk for wounding to protect the skin from wounding when possible.
Retention of bandages	Dressings are typically retained with rolled gauze and a retention stocking.
Cleanup	Cleanup and removal of old dressings, bandage packaging, needles, and so forth. Typically completed by a caregiver or nurse.
The above process of wound care can often take up to several hours to complete and is typically repeated daily or every other day for the life of the patient. The physical and emotional impact of EB-related wound care alone should not be underestimated.	

Sources: Bodan, 2016; Denyer, 2010; and Mauritz et al., 2019. EB = epidermolysis bullosa.



FIGURE 1. Sources of pediatric medical trauma in children with epidermolysis bullosa (EB). (a) Medical encounters (e.g., hospitalizations, procedures, clinic visits): skin erosion on the right ear incurred from repositioning of the jaw in the operating room in a child with severe recessive dystrophic EB. (b) Wound care (frequent, prolonged, painful, and anxiety provoking): often done by parents or caregivers in the home. Foot with blisters and keratoderma (from walking) requiring debridement in a child with severe EB simplex. (c) Community (including time spent with family and friends at home, at school, and in the community at large): wound resulting from a minor fall in a child with moderate recessive dystrophic EB.

Examples of the more unexpected sources of trauma for those with EB include (see Table 1)

- eating (e.g., painful mouth blisters or poor dentition making it difficult to chew; esophageal blistering making it difficult to swallow; choking and gagging because of food consistency, gastroesophageal reflux, or esophageal stricture; previous choking; Feijoo et al., 2011; Hubbard & Mayre-Chilton, 2015);
- walking (e.g., blistering and keratoderma from walking, EB-related neuropathic pain of the feet)
 - walking in the hallways at school (e.g., getting bumped into or stepped on, skin erosions or blistering from backpacks);
- time spent with friends and family (e.g., being held improperly can cause bullae or significant skin erosions; emotional impact of not being unable to keep up with peers or experiencing the physical consequences when they try to; Williams, Gannon, & Soon, 2011);
- sleeping (e.g., wounds adhering to clothing or sheets, corneal abrasions experienced during sleep making it difficult and painful to open the eyes); and
- injury from well-meaning healthcare providers (e.g., unintended shearing of the skin in the operating room, blistering from venipuncture; van Scheppingen, Lettinga, Duipmans, Maathuis, & Jonkman, 2008).

Although providers who care for children with EB are aware of the trauma related to wound care and surgical procedures, they may be less aware of the breadth and depth of “opportunities” for trauma that this population experiences. A deeper understanding of the daily life of children

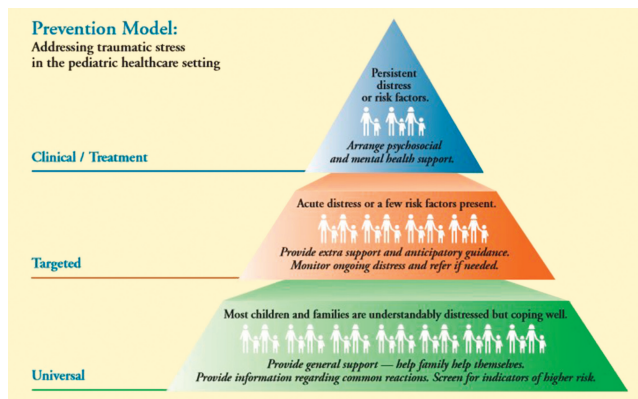


FIGURE 2. National Child Traumatic Stress Network (2014) prevention model of pediatric medical trauma.

and families impacted by EB may have the potential to improve care and outcomes for affected families. Interventions do not need to be prolonged or require excessive involvement by the healthcare team (see Figure 2 and Table 3). Families caring for children with the complex needs of EB are overwhelmed by numerous medical appointments and the time invested in providing care for their child (van Scheppingen et al., 2008). Intervening early with brief measures may be the best way to assist families who are not in immediate crisis. For many, acknowledging their

TABLE 3. Examples of Trauma-Informed Interventions for the Population With EB (Adapted From the NCTSN Model)

Intervention	Examples
Preventative measures	Pain management; education of families and local providers about how to limit distress and minimize trauma; connect patients with other families who have children with severe EB (e.g., camps, national EB conferences or meetings, or even social media connections); provide an understanding that some trauma is an inherent part of living with the condition.
Providing extra support	Identification of subclinical symptoms; short-term interventions; provide emotional support in the clinic setting.
Referrals	In the presence of persistent distress and/or negative impact on school, home refer for mental health intervention. Parents of school-age children may inquire about school-based counseling. Caregivers experiencing psychosocial distress may also be encouraged to seek care.

Sources: Goldschneider et al., 2014; Martin et al., 2019; NCTSN, 2014; van Scheppingen et al., 2008; and Williams et al., 2011. EB = epidermolysis bullosa; NCTSN = National Child Traumatic Stress Network.

hardships may be the needed intervention. For others, referrals to mental healthcare providers may be necessary.

TRAUMA IN CHILDREN WITH EB BEGINS AT BIRTH

Trauma for most with EB begins at birth with the initial days or weeks spent in the NICU. Because of the rarity of the condition, most families of newborns will find themselves in hospitals and NICUs with little to no experience in the delivery of care for this diagnosis. Many infants are born with no skin on many regions of the body and with large blisters that require extensive wound care (Ahmad & Bruckner, 2014). NICUs may discourage parents and medical staff from holding the infants to protect their skin (Bodan, 2016), which only adds to the stress of the encounter. Although most children will not remember their time in the NICU, there is some evidence to suggest that they may develop implicit memories of medical trauma, which may be implicitly triggered, as the child gets older (Gaensbauer & Jordan, 2009). This reinforces the need for providers to be aware that trauma (i.e., “trauma informed”) begins in the NICU (Bergeron, 2017; Lakatos, Matic, Carson, & Williams, 2019; Sanders & Hall, 2018). In fact, when NICU providers are trauma informed, they can increase connectedness between the infant and their parents (Sanders & Hall, 2018). This toxic stress exposure, which starts in the NICU, can be modified through social connection (Lakatos et al., 2019; Sanders & Hall, 2018).

Potentially toxic stressors for infants in the NICU include (Sanders & Hall, 2018)

- separation from parents;
- inconsistent caregivers;
- repeated painful procedures without environmental supports; and
- sensory environment that can overwhelm the immature brain.

THE IMPACT OF TRAUMA ON CHILDREN AND CHILDHOOD (NCTSN)

When children are exposed to repeated traumatic events in childhood, the responses can vary from positive, such as developing resilience, to negative including extensive, more serious and damaging impacts on the brain and nervous system (NCTSN, 2014). It is not uncommon for children who undergo acute medical events, because of illness or injury, to experience significant and persistent symptoms of posttraumatic stress (PTS; Bergeron, 2017). Below is a list of possible traumatic responses (NCTSN, 2014) and health outcomes linked to trauma exposure and pediatric medical trauma. These negative outcomes are thought to occur, whether or not the child meets diagnostic criteria of PTS syndrome or not (Marsac, Kassam-Adams, Delahanty, Widaman, & Barakat, 2014).

Related to acute pediatric medical trauma (Holbrook et al., 2005; Kahana, Feeny, Youngstrom, & Drotar, 2006; Mintzer et al., 2005; Zatzick et al., 2008):

- Decreased functional health
- Increased use of health services

Related to pediatric trauma exposure in general (NCTSN, 2014):

- Intense and ongoing emotional upset
- Symptoms of anxiety and depression
- Behavioral changes
- Difficulties with self-regulation
- Problems relating to others or forming attachments
- Regression
- Attention and academic difficulties
- Nightmares
- Difficulty sleeping and eating
- Somatic symptoms
- Drug and alcohol use
- Risky behavior
- Unhealthy sexual activity

Not all children exposed to trauma will develop traumatic stress, and there is value in understanding the protective attributes that build resilience, which include (NCTSN, 2014)

- severity of the event;
- proximity to the event;
- caregiver's reactions;
- prior history of trauma; and
- family and community factors.

Being mindful of the mental health and traumatic responses of children and families affected by EB is a means to improve the quality, and even potentially the quantity, of their lives. The important role of family and caregivers in helping to build resilience for those impacted by trauma exposure is notable and serves as a reminder of the importance of considering the health and well-being of parents and caregivers of children with EB.

TIC: WHAT DOES IT MEAN AND WHY DOES IT MATTER?

Trauma-informed healthcare providers are able to recognize that their patients may have been exposed to many different types of trauma and that they may need support and understanding as a result of these traumatic experiences (NCTSN, 2014; Substance Abuse and Mental Health Services Administration [SAMHSA], 2014). The trauma-informed provider also understands that children who have been traumatized can be retraumatized by well-meaning providers and caregivers (NCTSN, 2014). In essence, when one fully grasps the impact of trauma and, with intention and purpose, promotes protective factors to build resilience, they are providing trauma-informed healthcare (Earls, 2018). The trauma-informed provider can minimize trauma potential during medical care, provide anticipatory guidance, and identify children in distress, making referrals when and where appropriate (NCTSN, 2014). Trauma-informed providers need not only be mental health professionals but rather be any healthcare professional caring for the child with EB. With awareness and training, all members of the healthcare team have the potential to provide effective TIC.

The potential for providers to unintentionally retraumatize children with EB is particularly poignant. Because of the



FIGURE 3. Examples of skin damage caused unintentionally in the medical setting in children with epidermolysis bullosa (EB): (a) damage to the foot during blood draw, junctional EB; (b) damage to the neck during repositioning of airway in the operating room, recessive dystrophic EB; (c) blisters because of blood pressure cuff on a child's calf, junctional EB. With careful attention to care, the above wounds may have been avoided.

fragility of their skin, it is not uncommon for children with EB to endure unintentional traumatic injuries from healthcare providers (see Figure 3; van Scheppingen et al., 2008). Being aware of this type of medical trauma in the population with EB and taking sufficient time to communicate with the child and caregivers is a simple yet important place where many providers caring for this population with EB can begin.

An important component of providing TIC is the promotion of resiliency. Children with a trauma history, who are also described as resilient, tend to perform better in school and are better able to maintain control of their emotions and behaviors, compared with those who do not have resilient characteristics (Sege & Harper Browne, 2017). Resilience may be an innate characteristic for some but can also be cultivated by caregivers and healthcare providers. Although healthcare providers will not be able to change the EB reality for their patients, they have the opportunity to conceptualize the impact of wound care and other issues encountered by those affected, in a manner that could mediate the consequences of the frequent and repetitive trauma for those with EB as well as their caregivers (Martin et al., 2019).

RECOMMENDATIONS AND RESOURCES FOR PROVIDERS

Pediatric providers, of all subspecialties, caring for children with EB should consider the intermittent but persistent trauma endured by this population. Although some children living with EB will no doubt be well adjusted and thrive without intervention, all could benefit from the provision of TIC. Providers have the opportunity to limit retraumatization, provide short-term interventions, assess, and refer patients appropriately.

Formal training on integrating TIC into practice is an ideal goal; however, there are practical resources providers may

consider integrating into their practice that do not require a “formal” structure. These resources include both the NCTSN DEF protocol and the “Four Rs” as published by SAMHSA (2014). In addition, the Child Trauma Screening Questionnaire (CTSQ) may help predict which children with EB are at risk for developing persistent traumatic stress (Kenardy, Spence, & Macleod, 2006). Finally, DEBRA International has recently published clinical practice guidelines (CPGs) for the care of children and adults with EB, which may serve as a resource for all members of the interdisciplinary team (Martin et al., 2019).

DEF Protocol

The NCTSN developed the DEF protocol, which offers providers tangible ways to assess for and treat pediatric medical traumatic stress. The intention of the NCTSN was for providers to first address the ABCs of the child's health (airway, breathing, and circulation), followed by the trauma-informed DEFs (distress, emotional support, and family; NCTSN, 2014).

- Reduce Distress
 - Assess pain, fears, worries, and loss
 - EB considerations:
 - EB involves frequent and often chronic pain (Goldschneider et al., 2014; Pope et al., 2012). Despite the fact that children with EB have been living with pain since birth, many families could still benefit from new ideas regarding both pharmacologic and nonpharmacologic pain and stress management techniques.
 - Inquire about history of unintentional injury by medical providers (see Figure 3).
 - Recent deaths in the EB community that the child and/or family are coping with?
 - Do any of the fears, worries, or loss involve experiences at school such as bullying or isolation? (van Scheppingen et al., 2008; Williams et al., 2011)
 - How is wound care going for the child and family? Could they benefit from new strategies to limit stress, anxiety, and pain? (Mauritz et al., 2019; van Scheppingen et al., 2008)
- Promote Emotional support
 - What does the child need; what/who has helped the child in the past.
 - EB considerations:
 - Unless, and even if, there are EB experts available for procedures or surgeries, it may be beneficial for the EB parent/caregiver to be present to avoid unnecessary injury to the child (van Scheppingen et al., 2008).
 - Does the child have access to necessary emotional support? Are friends and family sufficient, or could they benefit from counseling at school or otherwise? Could they benefit from contact with other children of similar age who have EB (Martin et al., 2019; Williams et al., 2011)?
 - Remember the Family

- Remember to check in with siblings and caregivers, inquire about other potential home stressors (e.g., housing or financial).
 - EB consideration:
 - Caring for children with EB can be incredibly time consuming, stressful, and expensive, often making it difficult to be employed or enjoy typical recreational activities (Kirkorian et al., 2014; Mauritz et al., 2019; van Scheppingen et al., 2008).
 - Expenses such as wound care supplies are not always covered, or fully covered, by insurance and can be very expensive or even difficult to obtain (Kirkorian et al., 2014; Stevens, 2014).
 - Siblings may struggle with the extra attention received by the child with EB.
 - Parents or caregivers may themselves also develop psychological sequelae from observing medical procedures, wound care, or complications in the medical setting or at home (Martin et al., 2019; van Scheppingen et al., 2008).

The Four Rs

Providers may also consider the “Four Rs” of TIC as published by SAMHSA, which include the four principles of “realize,” “recognize,” “respond,” and “resisting retraumatization” when it comes to the provision of TIC (SAMHSA, 2014):

- Realize the prevalence and impact of trauma
 - EB considerations: Be familiar with the depth and breadth of situations in which children with EB may experience trauma (Table 1).
- Recognize signs and symptoms of trauma
 - EB considerations: an understanding that trauma begins at birth for many children and families with EB. Symptoms of trauma may include nightmares, increased startle response, increases in heart rate in response to a trigger, or depressive symptoms (Marsac et al., 2014). Anticipatory anxiety may also be present. Be mindful of signs of trauma among caregivers as well.
- Respond intentionally by integrating trauma knowledge into practice, policy, and procedures
 - EB considerations: Specific training and teams of providers identified to specialize working with children with EB and families may be beneficial in a pediatric medical system.
- Resist retraumatizing in the medical setting
 - EB considerations: Inquire about previous traumatic experiences within the healthcare system. Take parental and patient suggestions and experiences seriously; may require providers to be creative in the way they perform procedures/surgeries to avoid unintentional damage to the skin.

Child Trauma Screening Questionnaire

The CTSQ is a self-administered tool that may have value for predicting risk for persistent medical-related PTS (Kenardy

et al., 2006). The CTSQ is a 10-item questionnaire developed for children 7–16 years old who have experienced accidental injury. Although not diagnostic, the questionnaire can identify children at risk for developing posttraumatic stress 1 and 6 months after an injury (Kenardy et al., 2006). There are currently no data on the use of this questionnaire in the population with EB, but it has the potential to help clinicians better understand which patients with EB may require further intervention. The CTSQ is copyrighted but can be obtained by contacting the author j.kenardy@uq.edu.au).

DEBRA International Psychosocial Guidelines

The recently published CPGs include an extensive review of the literature as it relates to the psychosocial needs of those affected by EB (Martin et al., 2019). Despite the paucity of research on the psychosocial health for children with EB, these CPGs evaluated all available evidence, providing clinicians caring for children, adults, and families affected by EB with the most up-to-date source of recommendations for the psychosocial care of these individuals.

CONCLUSIONS

By framing wound care, and other common experiences of children living with EB, as a form of medical trauma, the hope is to bring to the forefront the profound impact living with EB has on patients and families. Children with moderate to severe forms of EB encounter trauma throughout any given day, and this trauma can come in many different forms, including physical and emotional. The frequent encounters with the medical system are all opportunities to improve the health and well-being of children with this complex and potentially life-threatening condition. Providers who understand the depth and breadth of experiences for those living with EB, and can care for them through a lens of being trauma informed, will potentially enhance the quality and perhaps even the quantity of life for children living with EB. Clinical research describing the short- and long-term impact of medical trauma among children and families affected by EB, along with any potential benefits of the provision of TIC, is warranted. ■

Acknowledgments

I would like to thank my daughter Clara, an EB warrior, who reminds me what it is to be strong and resilient in the face of adversity each and every day; the greater community with EB whose shared experiences helped to inspire the article; the many EB warriors and families who generously sent their encouragement and photos for this article; and my husband Francisco Bodán, MSW, for his support and feedback.

REFERENCES

- Adni, T., Martin, K., & Mudge, E. (2012). The psychosocial impact of chronic wounds on patients with severe epidermolysis bullosa. *Journal of Wound Care, 21*(11), 528. doi:10.12968/jowc.2012.21.11.528
- Ahmad, R. C., & Bruckner, A. L. (2014). A survey of epidermolysis bullosa care in the United States and Canada. *Pediatric Dermatology, 31*(2), 169–175. doi:10.1111/pde.12258

- Bergeron, M. (2017). Factors that support the use of child-parent psychotherapy as an intervention for child-parent dyads exposed to pediatric medical traumatic stress. *Clinical Social Work Journal*, 45(1), 77–87. doi:10.1007/s10615-016-0585-2
- Bodan, R. (2016). Epidermolysis bullosa: An insider's perspective to a rare genetic connective tissue disorder. *Journal of the Dermatology Nurses' Association*, 8(1), 46–56.
- Boeira, V. L., Souza, E. S., Rocha Bde, O., Oliveira, P. D., Oliveira Mde, F., Rêgo, V. R., & Follador, I. (2013). Inherited epidermolysis bullosa: Clinical and therapeutic aspects. *Anais Brasileiros de Dermatologia*, 88(2), 185–198.
- Denyer, J. E. (2010). Wound management for children with epidermolysis bullosa. *Dermatologic Clinics*, 28(2), 257–264. doi:10.1016/j.det.2010.01.002
- Dures, E., Morris, M., Gleeson, K., & Rumsey, N. (2011). The psychosocial impact of epidermolysis bullosa. *Qualitative Health Research*, 21(6), 771–782. doi:10.1177/1049732311400431
- Dystrophic Epidermolysis Bullosa Research Association. (2018). What is epidermolysis bullosa?. Retrieved from <http://www.debra.org/whatiseb>
- Earls, M. F. (2018). Trauma-informed primary care: Prevention, recognition, and promoting resilience. *North Carolina Medical Journal*, 79(2), 108–112. doi:10.18043/nmc.79.2.108
- El Hachem, M., Zambruno, G., Bourdon-Lanoy, E., Ciasulli, A., Buisson, C., Hadj-Rabia, S., ... Bodemer, C. (2014). Multicentre consensus recommendations for skin care in inherited epidermolysis bullosa. *Orphanet Journal of Rare Diseases*, 9(1), 76. doi:10.1186/1750-1172-9-76
- Espeleta, H. C., Sharkey, C. M., Bakula, D. M., Gamwell, K. L., Archer, C., Perez, M. N., ... Mullins, L. L. (2019). Adverse childhood experiences and chronic medical conditions: Emotion dysregulation as a mediator of adjustment. *Journal of Clinical Psychology in Medical Settings*. doi:10.1007/s10880-019-09639-x
- Feijoo, J. F., Bugallo, J., Limeres, J., Peñarrocha, D., Peñarrocha, M., & Diz, P. (2011). Inherited epidermolysis bullosa: An update and suggested dental care considerations. *Journal of the American Dental Association (1939)*, 142(9), 1017–1025. doi:10.14219/jada.archive.2011.0321
- Felitti, V. J., Anda, R. F., Nordenberg, D., Williamson, D. F., Spitz, A. M., Edwards, V., ... Marks, J. S. (2019). Relationship of childhood abuse and household dysfunction to many of the leading causes of death in adults: The adverse childhood experiences (ACE) study. *American Journal of Preventive Medicine*, 56(6), 774–786. doi:10.1016/j.amepre.2019.04.001
- Fine, J.-D., Bruckner-Tuderman, L., Eady, R. A. J., Bauer, E. A., Bauer, J. W., Has, C., ... Zambruno, G. (2014). Inherited epidermolysis bullosa: Updated recommendations on diagnosis and classification. *Journal of the American Academy of Dermatology*, 70(6), 1103–1126. doi:10.1016/j.jaad.2014.01.903
- Fine, J. D., Johnson, L. B., Weiner, M., & Suchindran, C. (2008). Cause-specific risks of childhood death in inherited epidermolysis bullosa. *The Journal of Pediatrics*, 152(2), 276–280. doi:10.1016/j.jpeds.2007.06.039
- Gaensbauer, T. J., & Jordan, L. (2009). Psychoanalytic perspectives on early trauma: Interviews with thirty analysts who treated an adult victim of a circumscribed trauma in early childhood. *Journal of the American Psychoanalytic Association*, 57(4), 947–977. doi:10.1177/0003065109342589
- Goldschneider, K. R., Good, J., Harrop, E., Lioffi, C., Lynch-Jordan, A., & Martinez, A. E., Dystrophic Epidermolysis Bullosa Research Association International (DEBRA International) (2014). Pain care for patients with epidermolysis bullosa: Best care practice guidelines. *BMC Medicine*, 12, 178. doi:10.1186/s12916-014-0178-2
- Holbrook, T. L., Hoyt, D. B., Coimbra, R., Potenza, B., Sise, M., & Anderson, J. P. (2005). Long-term posttraumatic stress disorder persists after major trauma in adolescents: New data on risk factors and functional outcome. *The Journal of Trauma*, 58(4), 764–769. doi:10.1097/01.ta.0000159247.48547.7d
- Hubbard, L. D., & Mayre-Chilton, K. (2015). Quality of life among adults with epidermolysis bullosa living with a gastrostomy tube since childhood. *Qualitative Health Research*, 25(3), 310–319. doi:10.1177/1049732314549029
- Kahana, S., Feeny, N., Youngstrom, E., & Drotar, D. (2006). Posttraumatic stress in youth experiencing illnesses and injuries: An exploratory meta-analysis. *Traumatology*, 12, 148–161.
- Kazak, A. E., Kassam-Adams, N., Schneider, S., Zelikovsky, N., Alderfer, M. A., & Rourke, M. (2006). An integrative model of pediatric medical traumatic stress. *Journal of Pediatric Psychology*, 31(4), 343–355. doi:10.1093/jpepsy/sj054
- Kenardy, J. A., Spence, S. H., & Macleod, A. C. (2006). Screening for post-traumatic stress disorder in children after accidental injury. *Pediatrics*, 118(3), 1002–1009. doi:10.1542/peds.2006-0406
- Kirkorian, A. Y., Weitz, N. A., Tloughan, B., & Morel, K. D. (2014). Evaluation of wound care options in patients with recessive dystrophic epidermolysis bullosa: A costly necessity. *Pediatric Dermatology*, 31(1), 33–37. doi:10.1111/pde.12243
- Lakatos, P. P., Matic, T., Carson, M., & Williams, M. E. (2019). Child-parent psychotherapy with infants hospitalized in the neonatal intensive care unit. *Journal of Clinical Psychology in Medical Settings*. doi:10.1007/s10880-019-09614-6
- Margari, F., Lecce, P. A., Santamato, W., Ventura, P., Sportelli, N., Annicchiarico, G., & Bonifazi, E. (2010). Psychiatric symptoms and quality of life in patients affected by epidermolysis bullosa. *Journal of Clinical Psychology in Medical Settings*, 17(4), 333–339. doi:10.1007/s10880-010-9205-4
- Marsac, M. L., Kassam-Adams, N., Delahanty, D. L., Widaman, K. F., & Barakat, L. P. (2014). Posttraumatic stress following acute medical trauma in children: A proposed model of bio-psycho-social processes during the peri-trauma period. *Clinical Child and Family Psychology Review*, 17(4), 399–411. doi:10.1007/s10567-014-0174-2
- Martin, K., Geuens, S., Asche, J. K., Bodan, R., Browne, F., Downe, A., ... Mayre-Chilton, K. M. (2019). Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: Evidence based guidelines. *Orphanet Journal of Rare Diseases*, 14(1), 133. doi:10.1186/s13023-019-1086-5
- Mauritz, P., Jonkman, M. F., Visser, S. S., Finkenauer, C., Duipmans, J. C., & Hagedoorn, M. (2019). Impact of painful wound care in epidermolysis bullosa during childhood: An interview study with adult patients and parents. *Acta Dermato-Venerologica*, 99(9), 783–788. doi:10.2340/00015555-3179
- Mintzer, L. L., Stuber, M. L., Seacord, D., Castaneda, M., Mesrkhani, V., & Glover, D. (2005). Traumatic stress symptoms in adolescent organ transplant recipients. *Pediatrics*, 115(6), 1640–1644. doi:10.1542/peds.2004-0118
- Murat-Sušić, S., Husar, K., Skerlev, M., Marinović, B., & Babić, I. (2011). Inherited epidermolysis bullosa—The spectrum of complications. *Acta Dermatovenerologica Croatica*, 19(4), 255–263.
- National Child Traumatic Stress Network. (2014). Pediatric medical traumatic stress toolkit for health care providers. Retrieved from https://www.nctsn.org/sites/default/files/resources/pediatric_toolkit_for_health_care_providers.pdf
- Pagliarello, C., & Tabolli, S. (2010). Factors affecting quality of life in epidermolysis bullosa. *Expert Review of Pharmacoeconomics & Outcomes Research*, 10(3), 329–338. doi:10.1586/erp.10.28
- Pope, E., Lara-Corrales, I., Mellerio, J., Martinez, A., Schultz, G., Burrell, R., ... Sibbald, G. (2012). A consensus approach to wound care in epidermolysis bullosa. *Journal of the American Academy of Dermatology*, 67(5), 904–917. doi:10.1016/j.jaad.2012.01.016
- Price, J., Kassam-Adams, N., Alderfer, M. A., Christofferson, J., & Kazak, A. E. (2016). Systematic review: A reevaluation and update of the integrative (trajectory) model of pediatric medical traumatic stress. *Journal of Pediatric Psychology*, 41(1), 86–97. doi:10.1093/jpepsy/sjv074
- Pynoos, R. S., Steinberg, A. M., Layne, C. M., Liang, L.-J., Vivrette, R. L., Briggs, E. C., ... Fairbank, J. (2014). Modeling constellations of trauma exposure in the National Child Traumatic Stress Network core data set. *Psychological Trauma: Theory, Research, Practice, and Policy*, 6(Suppl. 1), S9–S17. <https://doi-org.lib-proxy.fullerton.edu/10.1037/a0037767>
- Sanders, M. R., & Hall, S. L. (2018). Trauma-informed care in the newborn intensive care unit: Promoting safety, security and connectedness. *Journal of Perinatology*, 38(1), 3–10. doi:10.1038/jp.2017.124
- Schober-Flores, C. (2003). Epidermolysis bullosa: The challenges of wound care. *Dermatology Nursing*, 15(2), 135–138.
- Sege, R. D., & Harper Browne, C. (2017). Responding to ACEs with HOPE: Health outcomes from positive experiences. *Academic Pediatrics*, 17(7S), S79–S85. doi:10.1016/j.acap.2017.03.007
- Stevens, L. J. (2014). Access to wound dressings for patients living with epidermolysis bullosa—An Australian perspective. *International Wound Journal*, 11(5), 505–508. doi:10.1111/j.1742-481X.2012.01116.x
- Substance Abuse and Mental Health Services Administration. (2014). Substance Abuse and Mental Health Services Administration's concept of trauma and guidance for a trauma-informed approach (HHS Publication No. (SMA) 14-4884). Retrieved from <https://store.samhsa.gov/system/files/sma14-4884.pdf>
- Tabolli, S., Pagliarello, C., Uras, C., Di Pietro, C., Zambruno, G., Castiglia, D., ... Abeni, D. (2010). Family burden in epidermolysis bullosa is high independent of disease type/subtype. *Acta Dermato-Venerologica*, 90(6), 607–611. doi:10.2340/00015555-0947
- van Scheppingen, C., Lettinga, A. T., Duipmans, J. C., Maathuis, K. G., & Jonkman, M. F. (2008). The main problems of parents of a child with epidermolysis bullosa. *Qualitative Health Research*, 18(4), 545–556. doi:10.1177/1049732308315110
- Williams, E. F., Gannon, K., & Soon, K. (2011). The experiences of young people with epidermolysis bullosa simplex: A qualitative study. *Journal of Health Psychology*, 16(5), 701–710. doi:10.1177/1359105310387954
- Zatzick, D. F., Jurkovich, G. J., Fan, M. Y., Grossman, D., Russo, J., Katon, W., & Rivara, F. P. (2008). Association between posttraumatic stress and depressive symptoms and functional outcomes in adolescents followed up longitudinally after injury hospitalization. *Archives of Pediatrics & Adolescent Medicine*, 162(7), 642–648. doi:10.1001/archpedi.162.7.642