

5023

Mr. Chairman and members of the Committee, my name is Laura Morris and I live in Wethersfield, Connecticut. I write this letter in support of Bill Number 5023, An Act Requiring Health Insurance Coverage for Wound Care for Individuals With Epidermolysis Bullosa. I am the founder of the Epidermolysis Bullosa Support Foundation and a mother of a 25-year daughter with Epidermolysis Bullosa (EB). EB is a rare genetic skin disease where collagen is missing, or the body simply does not produce enough of it. Because the skin is missing this protein, blisters develop easily from the slightest pressure of clothing or a passing brush against another person or object. These blisters are treated like second or third degree burn and in the most severe forms of EB affects 80% to 90% of the body. There are three types of EB, EB Simplex, Junctional EB and Dystrophic EB, each with range from mild to severe and can require major adjustments in the lifestyle of both the EB patient and his or her family. The depth of the blister location within the skin layers defines these different subtypes. EB Simplex occurs in the outer layer of the skin; Junctional EB and Dystrophic EB occur in the basement membrane zone. In severe EB, blisters are not confined to the outer skin. They may develop inside the body, in such places as the linings of the mouth, esophagus, stomach, intestines, upper airway, bladder, and the genitals. Flowing is a summary of some of the characteristics signs of various forms of EB.

- EB Simplex (EBS) – A generalized form of EBS usually begins with blistering that is evident at birth or shortly afterward. In a localized, mild form called Weber-Cockayne, blisters rarely extend beyond the feet and hands. In some subtypes of EBS, the blisters occur over widespread areas of the body. Other signs may include thickened skin on the palms of the hands and soles of the feet; rough, thickened, or absent fingernails or toenails; and blistering of the soft tissues inside of the mouth. Less common signs include growth retardation; blister in the esophagus; anemia (a reduction in the red blood cells that carry oxygen to all parts of the body); scarring of the skin; and milia, which are small white skin cysts.
- Junctional EB (JEB) – This disease is usually severe. The most severe forms, large, ulcerated blisters on the face, trunk and legs can be life-threatening due to complicated infections and loss of body fluid that leads to severe dehydration. Blisters that affect the esophagus, upper airway, stomach, intestines, and the urogenital system also threaten survival. Other signs found in both severe and mild forms of JEB include rough and thickened fingernails and toenails; a thin appearance to the skin (called atrophic scarring); blisters on the scalp or loss of hair with scarring; malnutrition and anemia; growth retardation; involvement of the soft tissue inside the mouth and nose; and poorly formed tooth enamel.
- Dystrophic EB (DEB) – The dominant and recessive inherited forms of DEB have slightly different symptoms. In some dominant and mild recessive forms, blisters may appear only on the hands, feet, elbows and knees; nails usually are shaped differently; milia may appear on the skin of the trunk and limbs; and there may be involvement of the soft tissues, especially the esophagus. The more severe recessive form (RDEB) is characterized by blisters over large body surfaces, loss of nails or rough or thick nails, atrophic scarring, milia, itching, anemia and

growth retardation. Severe forms of recessive DEB (RDEB) also may lead to severe eye inflammation with erosion of the cornea, early loss of teeth due to tooth decay and blistering and scarring inside the mouth and gastrointestinal tract. IN most people with this form of EB, some or all the fingers and toes may fuse. Also, individuals with recessive DEB (RDEB) have a high risk of developing a form of skin cancer called squamous cell carcinoma. It primarily occurs on the hands and feet. The cancer may begin as early as the teenage years. It tends to grow and spread faster in people with EB than in those without the disease.

How Is Epidermolysis Bullosa Treated?

Persons with the mild forms of EB may not require extensive treatment. However, they should attempt to keep blisters from forming and prevent infection when blisters occur. Individuals with moderate and severe forms may have many complications and require psychological support along with attention to the care and protection of the skin and soft tissues. Patients, parents or others care providers should not feel that they must tackle all the complicated aspects of EB care alone. There are doctors, nurses, social workers, clergy members, psychologists, dietitians and patient and parent support groups that can assist with care and provide information and emotional support.

Caring for Blistered Skin

When blisters appear, the objectives of care are to reduce pain or discomfort, prevent excessive loss of body fluid, promote healing and prevent infection.

The doctor may prescribe a mild analgesic to prevent discomfort during changes of dressings (bandages). Dressing that are sticking to the skin may be removed by soaking them off in warm water. While daily cleansing may include a bath with mild soaps, it may be more comfortable to bathe in stages where small areas are cleaned at a time.

Blisters can become quite large and create a large wound when they break. Therefore, a medical professional will likely provide instructions on how to safely break a blister in its early stages while still leaving the top skin intact to cover the underlying reddened area. One technique is to pat the blister with an alcohol pad before popping it at the side with a sterile needle or other sterile tool. The fluid can then drain into sterile gauze that is used to dab the blister. After opening and draining, the doctor may suggest that an antibiotic ointment be applied to the area of the blister before covering it with a sterile, non-sticking bandage. To prevent irritation of the skin from tape, a bandage can be secured with a strip of gauze that is tied around it. In milder cases of EB or where areas are difficult to keep covered, the doctor may recommend leaving a punctured blister open to the air.

A moderately moist environment promotes healing, but heavy drainage from blister areas may further irritate the skin and an absorbent or foam dressing may be needed. There are also contact layer dressings where a mesh layer through which drainage can pass is placed on the wound and is topped by an outer absorbent layer. The doctor or other health care professional may recommend gauze or bandages that are soaked with petroleum jelly, glycerin, or moisturizing substances, or may suggest more extensive wound care bandages or products.

Treating Infection

The chances of skin infection can be reduced by good nutrition, which builds the body's defenses and promotes healing, and by careful skin care with clean hands and use of sterile materials. For added protection, the doctor may recommend antibiotic ointments and soaks.

Even in the presence of good care, it is possible for infection to develop. Signs of infection are redness and heat around an open area of skin, pus or a yellow drainage, excessive crusting on the wound surface, a red line or streak under the skin that spreads away from the blistered area, a wound that does not heal and/or fever or chills. The doctor may prescribe a specific soaking solution, an antibiotic ointment, or an oral antibiotic to reduce the growth of bacteria. Wounds that are not healing may be treated with a special wound covering or biologically developed skin.

Treating Nutritional Problems

Blisters that form in the mouth and esophagus in some people with EB are likely to cause difficulty in chewing and swallowing food and drinks. If breast or bottle feeding results in blisters, infants may be fed using a preemie nipple, a cleft palate nipple and eyedropper or a syringe. When the baby is old enough to take food, adding extra liquid to pureed food makes it easier to swallow. Soups, milk drinks, mashed potatoes; custards and puddings can be given to young children. However, food should never be served too hot. In the most severe forms of EB, a gastrointestinal tube is inserted and liquid supplements are given daily.

Surgical Treatment

Surgical treatment may be necessary in some forms of EB. Individuals with the severe forms of the autosomal recessive Dystrophic EB whose esophagus has been narrowed by scarring may require dilation of their esophagus for food to travel from the mouth to the stomach. Patients whose fingers or toes are fused together may require surgery to release them.

There is no cure for EB. The only treatment is proper wound care. This vital coverage acts as the skin for individuals with EB. Without it, infection and sepsis almost always ensues. These dressings are changed daily and sometimes twice daily to prevent infection. The parents and families of individuals with EB revolve

around day-to-day wound care. There is only ONE treatment for this condition: cover the chronic wounds with ointments and bandages. Unfortunately these supplies are outrageously expensive and some families pay as much as \$5,000 a month for these products. Which family can afford this? Many families are forced to skimp on their supplies; some have to lie about their income so they can apply for social programs. No family can afford such a bill unless they are independently wealthy.

Last year, Representative Morin, Representative O'Connor, my daughter Rachel and myself met with insurance industry leaders who supported and endorsed us.

There are approximately 50 individuals living with EB in Connecticut. Approximately 16 have the dominant dystrophic form of EB, 10 have the recessive dystrophic form of EB and 22 have the simplex form of EB.

Attached is an itemization of the cost of wound care coverage for my daughter, Rachel, who has the recessive form of EB.

I ask you to please carefully consider this bill. Thank you.

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Cost of Wound Care Coverage for a Person with Recessive Dystrophic Epidermolysis Bullosa

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| Telfa dressings (\$.05 per unit x 500 per monthj) | = \$ 250.00 |
| Mepilex dressing (\$11.99 per unit x 25 per month) | = \$1,199.00 |
| Flexicon bandage 6 inch (\$2.49 per unit x 100/month) | = \$ 249.00 |
| Flexicon bandage 4 inch (\$1.49 per unit x 48/month) | = \$ 73.00 |
| Guaze pads 4x4 (\$.20 per unit x 100/month) | = \$ 20.00 |
| Bactroban ointment (\$45.99 per tube x 16/month) | = \$ 736.00 |
| Micropore Tape (\$1.95 x 4/month) | = \$ 8.00 |
| Latex gloves (\$15.00 per box x 1 box/month) | = \$ 15.00 |
| 7 cases Ensure (\$62.48 per case x 7/month) | = \$ 38.00 |
| Enteral nutrition bags for Gtube (\$14.83 per unit x 30/month) | = \$ 445.00 |
| Bard Gtube connector (\$8.33 per tube x 30/month) | = \$ 250.00 * |
| | |
| Total Monthly | \$ 3,447.00 |
| | |
| Total Yearly | \$ 41,366.00 |

*Not covered under Medicaid