WHEN CHILDHOOD HURTS: EPIDERMOLYSIS BULLOSA

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CONFLICT OF INTEREST DISCLOSURE

• JENNY STYERS, RN-BC, MSN, CPNP-AC/PC
  • NOTHING TO DISCLOSE
• LYNN CLARK, RN, MS, CPNP-PC, AP-PMN
  • NOTHING TO DISCLOSE

OBJECTIVES

• IDENTIFY THE 4 TYPES OF EB
• RECOGNIZE IMPLICATIONS OF EB-ASSOCIATED PAIN THROUGHOUT CHILDHOOD
• IDENTIFY RECOMMENDED TREATMENT MODALITIES FOR EB-ASSOCIATED CHRONIC AND ACUTE PAIN
• STATE PRACTICAL APPLICATION OF TREATMENT OPTIONS FOR CHILDREN WITH EB
EPIDERMOLYSIS BULLOSA

• Significant blistering and erosions due to mechanical fragility of epithelial tissues
• Autosomal dominant or recessive inheritance
• Abnormal gene coding → altered structural protein formation → impaired integrity & function of skin and mucosa
• 4 main types of EB with many subtypes
• Incidence/occurrence
• No cure

EB DIAGNOSIS

• H&P including extensive family history
• “Onion skin” approach

TYPES OF EPIDERMOLYSIS BULLOSA: EB SIMPLEX (EBS)

• Characterized by blistering in the epidermis
• 75-85% of EB cases
• Localized or disseminated blisters
• Can occur on all skin & mucosal surfaces based on subtype
• Typically no major scarring but may have pigment changes, atrophy, milia
TYPES OF EPIDERMOLYSIS BULLOSA: JUNCTIONAL EB (JEB)

- Characterized by blisters within the basement membrane zone (BMZ)
- Localized or disseminated blisters
- Scarring common

TYPES OF EPIDERMOLYSIS BULLOSA: DYSTROPHIC EB (DEB)

- Characterized by blistering in the top layer of dermis
- Extreme dermis fragility, excessive scarring of skin & mucosa, nail changes
- Milia present
- Contractures and pseudosyndactyly
- Significant mucous membrane involvement

TYPES OF EPIDERMOLYSIS BULLOSA: KINDLER

- Characterized by blistering within multiple layers of epithelium + photosensitivity
- Generally presents as blisters to distal portions of limbs and ears
- Later develop photosensitivity, pokeweed, skin atrophy to UV-exposed areas
- Mucous membranes often affected
**MANIFESTATIONS AND COMPLICATIONS:**

**CUTANEOUS**
- Blisters to skin surfaces
- Impaired wound healing
- Skin atrophy
- Erosions, granulation tissue
- Scarring
- Pigment changes
- Hair loss/changes
- Nail changes
- Milia

**EXTRACUTANEOUS**
- Eyes: Corneal ulcerations, erosions, scars, conjunctival erosions
- Oropharynx: Blisters, tooth/ enamel changes, dental caries
- GI Tract: Dysphagia, reflux, strictures, impeded peristalsis, absorption issues, constipation, anal fissure, painful direction
- Respiratory Tract: Blisters, edema, granulation tissue/ scars, hoarseness, laryngeal stenosis, dyspnea, pneumonia, acute airway obstruction
- GU Tract: Dysuria, hematuria, stones, obstruction, vesicoureteral reflux, renal failure, renal HTN
- Metabolism: Nutrition deficiencies, increased caloric needs, FTT
- Other: Chronic anemia, cardiac abnormalities, osteopenia, osteoporosis, infections, pruritis, deconditioning

**EB PAIN THROUGHOUT CHILDBIRTH**
- Anticipatory pain
- Acute
- Chronic
- Psychosocial impact
EB PAIN THROUGHOUT CHILDHOOD: ANTICIPATORY PAIN

- Prevention of situations that can lead to trauma to the skin or exacerbate other pain-inducing complications is the primary goal.
- Anxiety can be significant, so anxiolytics are important.
- Using appropriate dressings, wraps, padding, and finding activities that are “safe” but fun is a priority.
- Treating anxiety can significantly reduce the amount of opioids required to complete tasks.

EB PAIN THROUGHOUT CHILDHOOD: ACUTE PAIN

- Occurrence
  - Before, during, and after blisters
  - Friction
  - Walking
  - Heat
  - Sweating
  - Trauma
  - Dressing changes
  - Seasonal

EB PAIN THROUGHOUT CHILDHOOD: CHRONIC PAIN

- Neuropathic pain
- Pain hypersensitivity and amplification
- Neurochemicals
- Epigenetics
- Other factors
  - Descending pain modulatory system (DPMS)
  - PRRTs
EB PAIN THROUGHOUT CHILDHOOD
PSYCHOSOCIAL IMPACT

- Quality of life
- Social life
- Mental health
- School/Work
- Financial burden

SECONDARY COMPLICATIONS THAT HAVE PAIN IMPLICATIONS

- Pruritus
- Infection
- Nutritional deficiencies
- Osteoporosis

MANAGEMENT OF EB-ASSOCIATED ACUTE PAIN

- Acute trauma - wounds
- Dental pain
- Blistering of esophagus
- Corneal ulcerations
- Defecation/Urination
- Procedural pain
- Psychological
MANAGEMENT OF EB-ASSOCIATED
CHRONIC PAIN

- DAILY PAIN
- CHRONIC WOUNDS
- PRURITUS
- CONTRACTURES
- OSTEOPOROSIS
- PAIN FROM DYSPHASIA
- DECONDITIONING
- PSYCHOLOGICAL

PRACTICAL APPLICATION OF TREATMENT OPTIONS
FOR CHILDREN WITH EB

- EARLY RECOGNITION OF EB DIAGNOSIS
- EARLY RECOGNITION AND TREATMENT OF PAIN
- PATIENT AND PARENT EDUCATION
- COMMUNITY RESOURCES
  - DYSRHEINIC EB RESEARCH ASSOCIATION OF AMERICA WWW.DEBRA.ORG
  - FACEBOOK GROUPS
  - NATIONAL CAMPS
    - WWW.EBKIDS.ORG
    - WWW.EBNURSE.ORG

MULTIDISCIPLINARY TEAM

ON-GOING
- PSY
- EB CENTERS
- DERMATOLOGY
- WOUND CARE
- PAIN MANAGEMENT
- PSYCHOLOGY/PSYCHIATRY

AS NEEDED
- GENETICS
- DENTISTS
- OPHTHALMOLOGISTS
- SOCIAL WORK
- PT / OT / SPEECH
- PALLiative CARE
SPECIAL CONSIDERATIONS

• SCHOOL ACCOMMODATIONS
• SOCIAL ACTIVITIES
• SUPPORT / RESEARCH
• TRANSITION TO ADULTHOOD