Occupational Therapy and Physiotherapy for patients with Epidermolysis Bullosa
Presenters

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Objective

• To increase the awareness of the role Occupational Therapy and Physiotherapy have in improving the overall function of the patient with Epidermolysis Bullosa

• Review adaptations for functional and school related tasks for the patient with Epidermolysis Bullosa

• To raise awareness of how functional and therapeutic needs change with age in the patient with Epidermolysis Bullosa

• To review Hand Therapy Management of the adult Hand
Children and Adults with EB are at increased risk for debilitation, limited range of motion, and weakness due to blistering.

What can therapy offer?

- Evaluation
- Intervention
- Adaptations and modifications
- Outcome monitoring
Occupational & Physical Therapy Evaluation

Daily Activities: ADLs, school, sports, hobbies

Is there a delay in development or a risk for delay?

Family Centered Care & Patient Goals

Are there limitations due to loss of motion or blistering?
Questions in regards to the Neonate

**Therapist**
- Skin integrity
- How much damage has taken place?
- Where is the damage?
- How about the oral mucosa? Are there blisters in the mouth?
- How is the neonate feeding? Reflux? Aspiration? Breastfeeding vs normal bottle vs Haberman Feeder

**Family**
- How can I help my baby?
- Is it okay to hold him?
- Is it okay to give him a pacifier?
- Can I breastfeed? Should I breastfeed?
What are we evaluating?

- Neonate: birth trauma
- Infant: developmental milestones
- Toddler: developmental milestones
- School Age: peer interaction
- Adolescent: puberty, transition to independence
- Adult: work, hobbies
Questions in regards to the Infant

**Therapist**

- Can the infant move through full ranges of motion? If not, where is she limited?
- Is the infant achieving milestones? If not, where are the limitations and what can we do to assist?
- How can we protect the skin?

**Family**

- Is my child reaching developmental milestones appropriately?
- How can I protect my child’s skin?
- Is it okay to provide tummy time for my child?
**Questions in regards to the toddler**

**Therapist**

- Is the toddler achieving milestones appropriately?
- How is the toddler bandaged?
- Is the toddler participating in age appropriate activities with peers?

**Family**

- Is my child doing all he should be?
- What can I do to assist with protecting his skin?
Questions in regards to the preschooler

**Therapist**

• Are they performing age appropriate activities with their peers?

• Are they achieving appropriate developmental milestones?

**Family**

• How can I protect my child from being bumped or hit during preschool activities?
Questions in regards to School age

**Therapist**
- Is the child performing age appropriate activities?
- Is she able to sit in classroom without pain?
- Is she participating in gym class with her peers?
- Is she participating in recess?

**Family**
- Should I get a wheelchair for my child?
- Would my child benefit from an orthopedic surgery?
Adolescent and Adulthood issues

**Therapist**
- Physical limitations
- Activities of Daily Living

**Family/self**
- Difficulties with everyday activities
- Need for surgery?

**Most Common Orthopedic Surgeries**
- Hand/Finger releases
- Hamstring releases
- Ankle fusions
- Toe surgeries
Intervention

• *With neonates, infants, toddlers, preschoolers:*
  – Parent and child education to help promote normal development

• *With all ages:*
  – Gentle stretches to help maximize movement
  – Strengthening activities to help increase endurance and function
  – Modification of activities and/or adaptive equipment to encourage independent and successful performance of activities of daily living
  – Splinting to help preserve range of motion and function
Early Development
Why is belly time so important?

• Helps to build strength as baby learns to lift head, reach arms, and roll over.
• These skills lead to sitting, crawling, walking.
• Helps encourage sensory exploration. This is how a baby learns about the environment.

Modifications
• Baby can be placed on belly on parents chest if there is too much discomfort on the floor.
• Parents can pad knees to help reduce potential blistering
Importance of Typical Childhood Activities

- Children learn and develop strength through play and through their engagement in daily activities.
- It is crucial to allow them to participate in typical play activities in the safest manner possible.
Typical activities that help to strengthen muscles

- Crawling
- Walking
- Push toys
- Self care
- Riding a bike
- Non contact sports
- Wii Fit and/or Kinect
- Dance
- Yoga
What can I do to help my child succeed?

- Wrap with the appropriate protection. Do not overwrap so the child cannot move.
- Treat them as a child without EB while protecting the skin.
- Provide touch and allow them to touch and feel different textures.
- Promote independence rather than dependence
Adaptations & Modifications

• Preserving strength and range of motion are always important. However, sometimes the activity or the environment can be modified for greater success or for improved quality of life.
• Many patients with EB benefit from daily adaptations and modifications in the areas of self care, school/work, and play/leisure.
• The following slides outline commonly used adaptations
Adaptations & Modifications

Self Care

• Adapt Clothing fasteners with magnets, velcro closures, sliding hooks
• Cutting buttonholes larger
• Seamless clothing and socks
• Use zipper pulls or loops
• Explore shoes such as Crocs, Ugg, Pedors
Adaptations & Modifications

School

- Use soft pencil grips and/or mechanical pencils and easy glide pens to minimize pressure
- Ask for an extra set of books in each room
- May need extra time for written work and assignments
- Explore using a cushion on chair during class as needed
Adaptations & Modifications

School Technology - Computers and tablets are often used in conjunction with or instead of handwriting

- Live Scribe Pen – writes and records speaker while writing
- Speak to Type software: Dragon Naturally Speaking – can work with any computer or tablet, WordQ + Speak Q, Co:Writer6
- Speak to Type Apps: Write and Say, Dragon Dictation, PaperPort,
- Word Prediction Apps: Abilipad, Cowriter6
Splinting

- Commonly used to help preserve range of motion
- Limited research on effectiveness with patients with EB, particularly prior to hand release surgery
- Most effective when combined with regular stretching
- Many patients prefer to wrap hands than wear a splint
Splinting

**Benefits**
- May help to limit loss of motion
- Variety of splinting options based on the patients needs

**Challenges**
- Heat and moisture
- Friction
- Compliance – splints can be an uncomfortable addition to an already uncomfortable person
Outcome monitoring

• Are we helping to improve quality of life?
• Has range of motion or strength changed?
• Have previous goals been met? Have the patient or family goals changed?
• Is the patient/family able to participate in meaningful activities?
Outcome monitoring

• Patient and family goals change throughout the lifespan, as do therapeutic goals
• Therapists can be a useful resource at each stage in life, however ongoing therapy is not always indicated
• It is the role of the therapist to help patients and families consider their goals and the importance and relevance of therapy at each stage in life
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Thank you Rick Guidotti and families!