

Palliative and End-of-Life Care for People Living with Epidermolysis Bullosa

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REVIEW

Open Access

Consensus-based guidelines for the provision of palliative and end-of-life care for people living with epidermolysis bullosa



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- K.M.C. declared a potential conflict from her professional work coordinating guidelines for DEBRA International.
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Coming Soon. . .



PALLIATIVE & END-OF-LIFE CARE
Information for people with epidermolysis bullosa
(EB), their families, and carers



Definitions



What is Palliative Care?

“An approach that **improves the quality of life of patients** (adults and children) **and their families** who are facing problems associated with life-threatening illness. It **prevents and relieves suffering** through the **early identification, correct assessment and treatment** of pain and other problems, whether **physical, psychosocial, or spiritual.**”

World Health Organization, 2020

So, what is “Hospice” or “End-of-Life” Care?

- Comprehensive comfort care and family support for a person who has a serious illness **AND** is approaching the end of their life.
- Attempts at cure are stopped
- No attempts are made to prevent or accelerate the dying process
- Treatment focuses on
 - quality of life,
 - symptom management,
 - comfort, and
 - dignity.

How About “Comfort Care”?

- Provided when the end of life is near.
- Focuses on symptom control, pain relief, and quality of life.
- Goal is to control pain and other symptoms so the patient can be *as comfortable as possible*.

Palliative Care

Hospice or
End-of-Life
Care



Comfort Care



Therefore. . .

All EB-related care is palliative!





Methods



Methods

- Questionnaire distributed world-wide that sought opinions related to the provision of palliative care in the EB community.
- Three broad clinical questions:
 1. What are the best practices to manage the symptoms of, and improve survivorship for, EB over the lifespan?
 2. Are there recommendations and interventions to improve mental health; quality of life; and overall life satisfaction with respect to cultural, religious, and other belief-systems of people living with EB?
 3. What are best practices to identify individuals who are grieving as a result of EB and to assist them in improving their ability to cope with that grief?

Methods

Six clinical outcomes:

1. Symptom management
2. Survivorship
3. Mental health/Well-being
4. Quality of life
5. Life satisfaction
6. Grief

Methods

- 1,405 articles identified using EMBASE, PubMed, & CINAHL
- Declined if:
 - Lay publications (newspapers, magazines, blog posts, etc.)
 - Did not meet the established criteria
 - Did not directly address clinical outcomes
 - If associated with, or were better addressed by, another CPG
- Used 85 articles
 - Each article appraised twice
 - Graded with GRADE methodology
- Used AGREE II tool on our own manuscript to increase the quality
- Final manuscript reviewed by 8 independent experts in EB before submitting for publication. Reviewed by 2 journal reviewers.



Recommendations

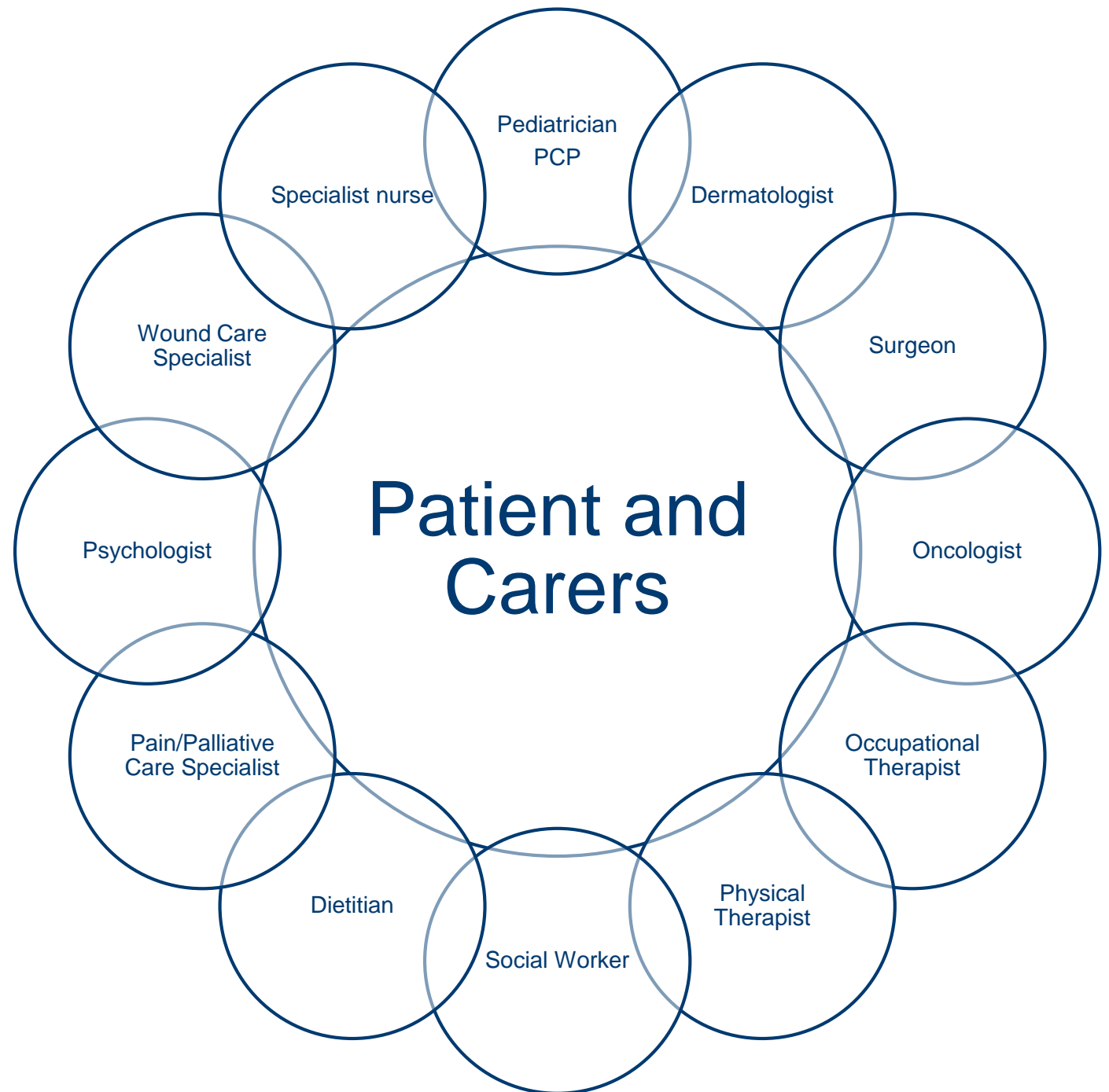


Recommendations

- Formulated 55 recommendations
 - Unanimously agreed upon
 - Used GRADE tool, expert opinion, and panel consensus
- Recommendation strength strongly influenced by lack of high-quality evidence and expert panel decision-making, which accounts for observable gaps between evidence levels and recommendation strength.
- Evidence level very low for all recommendations.

Key Recommendation

All care should be **interdisciplinary** and should include:





Geneticist



Wound care nurse



General Practitioner



Pediatrician



Dermatologist



Gastroenterologist



Speech therapist



Psychologist



Pain specialist



Person living with EB and their Carers



Surgeon



Otolaryngologist



Podiatry care



Palliative care specialist



Oncologist



Cardiologist



Ophthalmologist



Occupational therapist



Nephrologist



Anaesthetist



Specialist Nurse



Nutritionist/dietitian



Social care



Dentist



Physiotherapist



Endocrinologist

Key Recommendations – Question 1: Symptom Management and Survivorship

- Provide adequate pain and itch control
- Focus on achievable goals over the *entire* lifetime, with the goal of managing physical and/or emotional suffering while respecting the autonomy and individuality of each person, and providing psychoeducation to the person living with EB, their family, and carers in a way they can fully understand.
- Fully inform the patient of all their treatment options (including no treatment) in an age-appropriate manner
- NG/NJ tubes are not recommended because of the facial damage they cause
- Focus exclusively on comfort care for severe Junctional EB
- Palliative sedation may be considered to relieve refractory symptoms at EoL
- Providers should use self-care practices

Key Recommendations – Question 2: Mental Health, QoL, and Life Satisfaction

- Consider the whole person living with EB rather than only their visible lesions
- As clinical severity increases so does the need for a holistic treatment approach
- People living with EB must have the same opportunities to feel useful and to be a contributing member of society just like those without EB
- Address the mental health of parents, carers, and providers
- Family members need long-term support
- Families and carers may need supportive care following bereavement

Key Recommendations – Question 3: Grief

- Provide psychosocial support for parents to address the trauma unique to parents of a child with EB
- Provide a strong support system throughout the dying process and beyond
- Health care providers should offer ongoing bereavement support following death
- Allow health care providers to attend funerals of their patients if allowed by the family



Clinical Implications



Clinical Implications

This evidence-based guideline describes ways to:

- optimize and standardize interdisciplinary care
- to reduce suffering, and
- improve comfort and overall quality of life



Future Research



Future Research

- Develop best care practices specific to:
 - Needs at end-of-life
 - EB subtype
 - Age
 - Areas with limited access to an interdisciplinary team and/or medical resources
- Evaluate generalizability of literature related to people with other potentially life-limiting conditions to people living with EB
- Examine how beliefs and regulations affect provision/reception of palliative care services within the EB population globally and during individual medical visits
- Explore the experience of grief in people living with EB across cultures and ethnicities
- Develop treatments/skills to reduce pain/itch without increasing side-effects
- Explore the challenges/process of, and bioethics related to, medical futility and the decisions to provide/alter/withhold aspects of care, especially at EoL, including terminating life-sustaining therapies as well as using comfort care, palliative sedation, and the Groningen Protocol/euthanasia, especially in cases of the most severe subtypes of EB

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Thank you!

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