Pregnancy, childbirth and aftercare in EB

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EB Clinet - October 2024

I declare that I have been a consultant, lecturer and/or researcher for:

AbbVie, Amryt Pharma/Chiesi Pharma, Avicanna Inc., Carnot, Eucerin, Galderma, Gencell pharma, Leo pharma, Novartis, Pfizer, Pierre Fabre, Roemmers, Sanofi

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GUIDELINES

BJD
British Journal of Dermatology

Recommendations on pregnancy, childbirth and aftercare in epidermolysis bullosa: a consensus-based guideline*

D.T. Greenblatt,[1] E. Pillay,[1] K. Snelson,[1] R. Saad,[2] M. Torres Pradilla,[3] S. Widhiati,[4] A. Diem,[5] C. Knight,[1] K. Thompson,[6] N. Azzopardi,[7] M. Werkentoft,[8] Z. Moore,[9] D. Patton,[9] K.M. Mayre-Chilton,[1,10] D.F. Murrell[11] and J.E. Mellerio[1]

2018-2020

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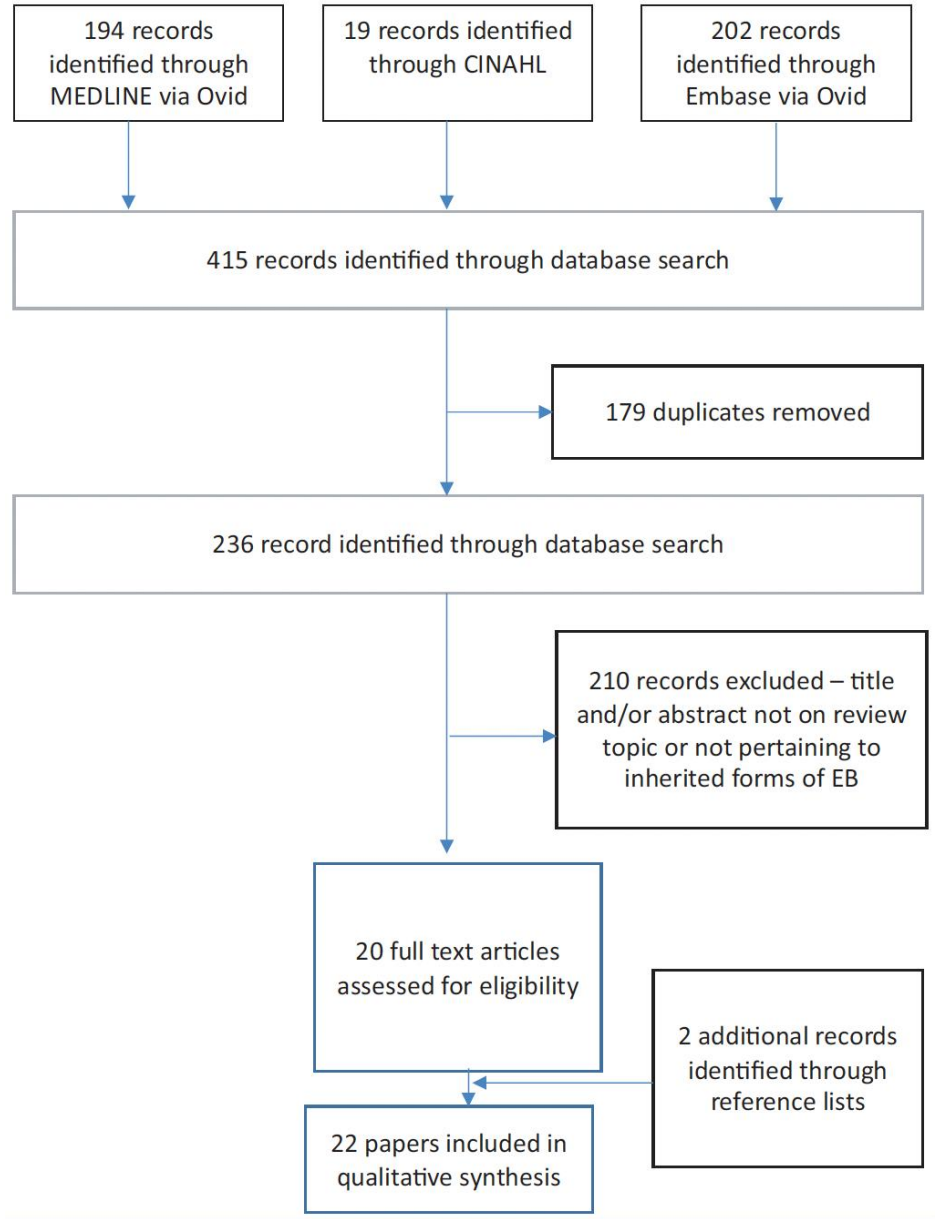
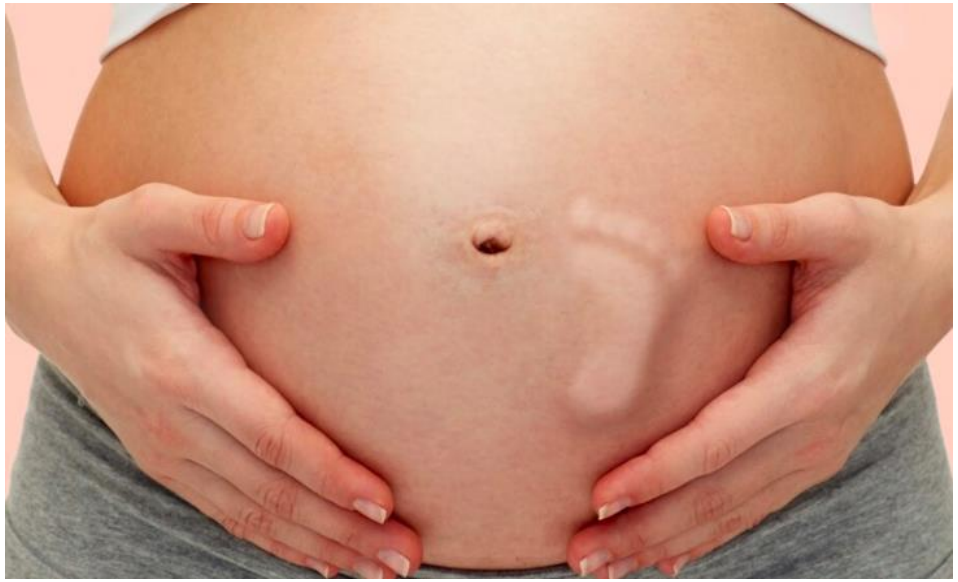


Preconception and antenatal management, fertility is not typically affected in milder forms of EB. However severe forms of EB may have an impact on ovulation.

Discuss and evaluate vulvovaginal manifestations.



Greenbalt DT et al, BJD 2021

1. Preconception and antenatal management

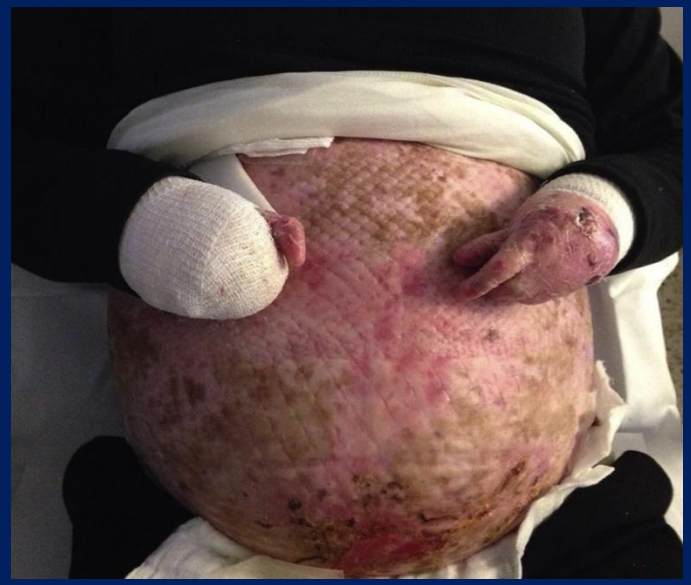
a. Diagnostics, genetic counseling and prenatal testing, offer standard preconception care to women and provide genetic counseling (dominant/recessive pattern)
Couples may wish for prenatal/preimplantation genetic testing.

b. Optimizing health pre-pregnancy, supplementing folate, managing iron and vitamin D levels; and supplementing zinc and selenium, if needed.

c. Medication review, review ALL active and OTC medicines and supplements.

Greenbalt DT et al, BJD 2021

1. Preconception and antenatal management



d. Physiological changes in pregnancy, rising levels of human chorionic gonadotropin in early pregnancy is associated with morning sickness, reflux and heartburn symptoms.
Monitor and treat nausea
Assess and manage constipation and gastric symptoms

e. Clinical examinations and antenatal visits, engage an MDT, specially for patients with complex forms of EB.
Perform standard antenatal investigations according to local guidelines: padding, non-adhesive tapes, lubrication.
Ultrasound can be performed safely

Greenbalt DT et al, BJD 2021
de Lucas R et al, Actas 2017

1. Preconception and antenatal management

f. Planning delivery, including anesthetic assessment, encourage women with EB to prepare a birth plan. Individualized approach, benefits and risks of all options should be discussed.
Having a diagnosis of EB in NOT a contraindication to vaginal birth.

Vaginal birth preferred mode for all EB subtypes.
Except: obstetric indication, extensive genital blisters/wounds, vaginal stenosis or breech presentation at term.

Prenatal anesthetic assessment on woman with known difficult airway, microstomia and wounds on lower back.

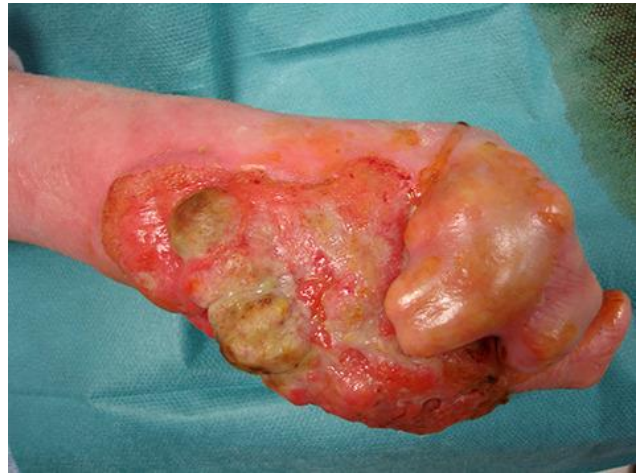
Greenbalt DT et al, BJD 2021

1. Preconception and antenatal management

g. Longer-term planning, plans for bringing home the newborn as soon as possible.
Considering maternal hand function and mobility, need for physiotherapy and review physical – emotional support.

Discuss breast- or bottle-feeding plans,

Offer ongoing routine EB skin monitoring and surveillance for skin cancer.



Greenbalt DT et al, BJD 2021
Moreira A et al, Ox Med Cas Repo 2020

2. Labour and management of delivery

a. Advance preparations, “dressing pack” including non adherent dressing and medical adhesive remover spray (MARS)

Patient “card”: EB subtype, delivery plan and contact details of MDT

b. Engagement of neonatal team, possibility for the baby to be born with EB. *handle with extreme care, avoid rubbing, lubricate catheters if needed.

Omaira (62) & Loraine (10)
DDEB

Greenbalt DT et al, BJD 2021

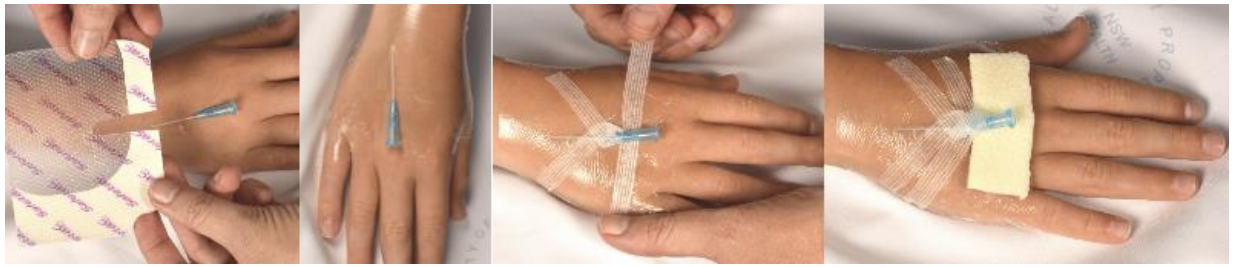
2. Labour and management of delivery

c. Monitoring and skincare during labor and delivery, gently examination and extra lubrication: gloves, speculum, etc.

d. Skincare management, inform staff about skin fragility, extra padding (table, BP cuffs, etc), avoid prolonged pressure/immobility, encourage self-positioning.

e. Catheterization, avoid unnecessary urinary catheterization.

f. Cannulation and securing IV lines, ultrasound-guided cannulation, if available, specially if access is difficult. Secured with low-adherent film.

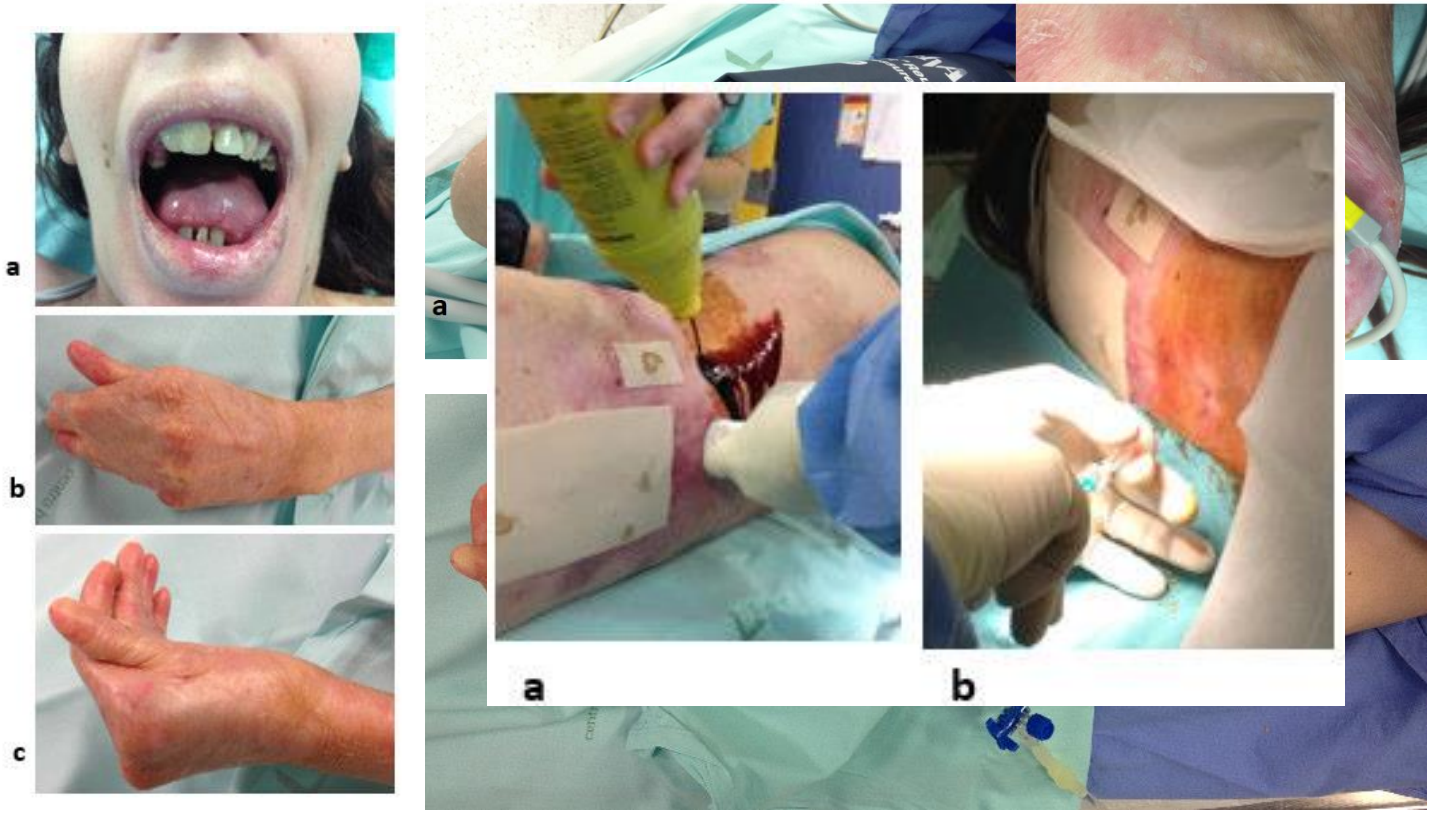


Greenbalt DT et al, BJD 2021

2. Labour and management of delivery

g. Analgesia and anesthetic management, follow the usual obstetric practice. If c-section is planned regional anesthesia should be offered. Skin preparation with care.

Extra precaution with general anesthesia in severe patients due to comorbidities.



Greenbalt DT et al, BJD 2021
Lemos P et al, IJOA 2017

2. Labour and management of delivery

h. Labor and birth setting, consider induction of labor if needed, is NOT contraindicated. Extra lubrication and gently as possible

Hydrotherapy during labor and water birth may be considered**.

Avoid instrumental delivery

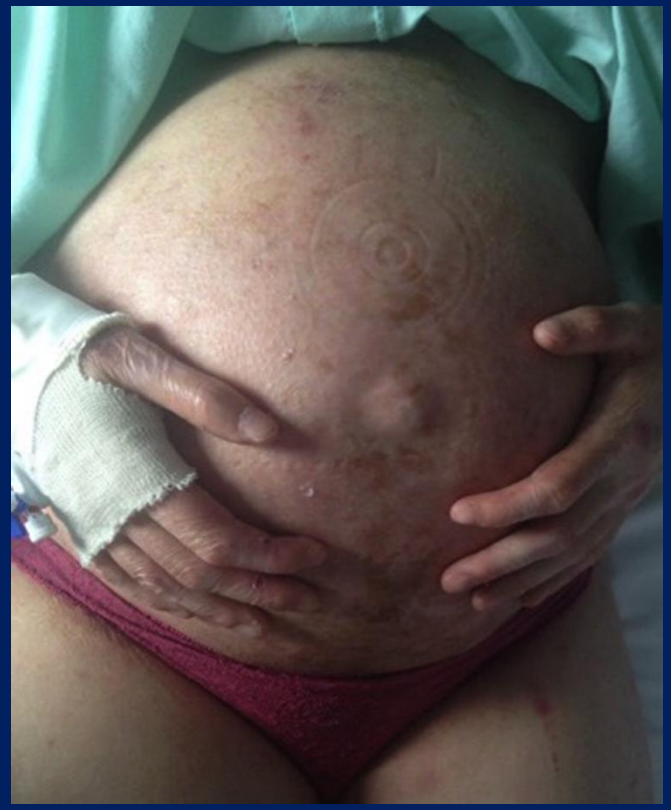
Cesarean section, if indicated, subcuticular sutures are acceptable

Episiotomies and tears heal well in EB

Avoid the use of compression stockings

Greenbalt DT et al, BJD 2021

3. Postnatal care and management



a. Perineal care, nonadherent dressings and soft sanitary pads. episiotomies and perineal lacerations tend to heal well in women with EB

b. Care of cesarean section wound, nonadherent dressings and MARS for C-section wound. Generally, heal well, blisters and erosions occasionally occurs.

c. Prevention of venous thrombosis, avoid compression stockings. EB do not represent higher risk of thromboembolism. Women at high risk of thromboembolism event benefit from low-molecular-weight heparin

Greenbalt DT et al, BJD 2021
de Lucas R et al, Actas 2017

3. Postnatal care and management

d. Infant feeding, **Asses factors such as the woman´s EB subtype, social support structures, condition of wounds on breasts or hands, and pain management.

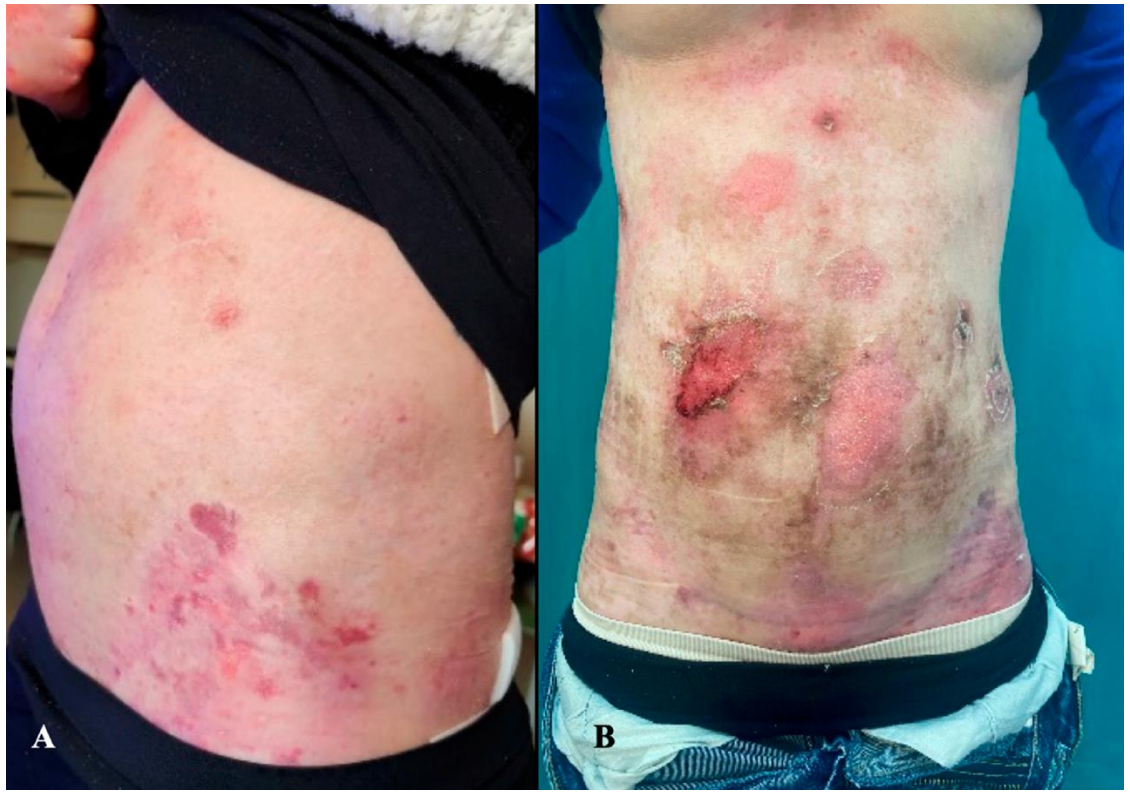
e. Breast feeding and breast care, open discussion antenatally. Evaluate wounds and possibility of new blisters.
Assist with positioning of the baby and adequate latching on to the areola. Well lubricated nipple shields may be useful.

f. Formula and mixed feeding, plan support for women with pseudosyndactyly who may need assistance with the preparation formula.
Included in the postnatal care plan if chosen.

Greenbalt DT et al, BJD 2021

3. Postnatal care and management

g. Planning discharge, women with EB do not need longer post-natal hospital admission.
May require physical aids in severe patients.
Engagement with support networks.
Screen for mood disorders
Schedule follow-ups post partum



Greenbalt DT et al, BJD 2021
Nicoli P et al, Diseases 2024

Women with EB should be encouraged in their reproductive choices.

With the appropriate genetic counselling, and a planned approach to care, positive pregnancy experiences and outcomes for mothers with EB and their babies can be achieved.

Muchas gracias!

www.debracolombia.org

15th Anniversary

Fundación Universitaria de Ciencias de la Salud
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Acreditación Institucional en Alta Calidad
Resolución 013601 de 2019 MEN Vigencia 4 años.

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