

Care of the woman with EB during pregnancy and childbirth

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DEBRA is the national charity that supports people living and working with Epidermolysis Bullosa (EB) – a genetic condition which causes the skin to blister and shear at the slightest friction or even spontaneously.

With grateful thanks to those people with EB who have shared their experiences of pregnancy and childbirth with the nursing team.

Who is this booklet intended for?

This booklet is for health care staff working in obstetrics that find themselves caring for women with epidermolysis bullosa. The information given is mainly regarding the nursing and midwifery care of women with the severer forms of EB, usually recessive dystrophic EB, non-Herlitz Junctional EB and dominant dystrophic EB. The information about skin care may also be helpful when looking after a woman with EB Simplex. However, your patient will be able to tell you of the extent of their problems and where caution is required. The Golden Rule in caring for someone with EB is **ASK THE PATIENT**.

What is EB?

The term EB refers to a group of genetically transmitted blistering disorders (Wojnarowska 1998). The common factor is the tendency of the skin and some internal mucosa to separate and form blisters and wounds. This tendency to separation occurs with minimal friction and trauma. There is a wide variation in genetic mutations causing this condition and EB is an umbrella term for a number of different conditions of widely varying degrees of severity.

There are three main forms of EB, as follows:

1. EB Simplex (EBS)

In EBS the fault lies within the scaffolding of the epidermis, Although the fragility extends all over the body, in the overwhelming majority of cases blistering is mainly confined to the hands and feet i.e. those areas subject to most trauma. Occasionally the blistering is more generalised and occurs in areas of friction e.g. waistbands, bra straps etc. This is a dominantly inherited condition (apart from very rare exceptions) and your patient, or their partner will normally have a long family history of the disorder. Occasionally the affected parent will not have a family history of the disorder This is the most common form of EB (Fine 1999)

2. Dystrophic EB (DEB)

In DEB the fragility is caused by absence of diminished anchoring fibrils as a result of a mutation to the collagen V11 gene. DEB can be inherited either recessively or dominantly. The anchoring fibrils attach the epidermis to the dermis. If they are deficient or absent the resulting skin fragility can be extreme. However, there is a wide spectrum of severity of DEB, with the recessive form tending to be more severe. The fragility in DEB can extend to the:

- a) Mouth and oro-pharynx
- b) Oesophagus
- c) Anal margins
- d) Urethral meatus and genital region
- e) Conjunctiva of the eye

In addition, wounds heal with scarring, which can in severe cases, lead to loss of functional digits as they are cocooned in mittens of skin. If this is a problem, surgery to separate the fingers is offered. However, as this is needed repeatedly, some individuals with EB prefer to manage without surgery. (Fine et al 1999)

3. Junctional EB (JEB)

In JEB the affected structures vary but the effect is to cause separation of the skin at the level of the lamina lucida of the basement membrane zone. It is important to note that there are 2 major forms of JEB-Herlitz Junctional EB, where the affected babies usually die in infancy, and non-Herlitz Junctional EB that is compatible with a near normal life span. JEB is always inherited as a recessive condition. In non-Herlitz Junctional EB the main effects upon the individual can be:

- a) Chronic non-healing ulcers
 - b) Alopecia
 - c) Hypoplasia of the dental enamel
 - d) Fragility of the genito-urinary tract
- (Fine 1999)

Treatment of all forms of EB

1. Prevention of new blistering
2. No adhesion, no friction
3. Bursting of new blisters to halt spread
4. Protection of the individual from infection
5. Application of appropriate dressings to aid wound healing
6. Treatment of pain

Additional treatment in the severe forms of EB

6. Surgical correction of deformities of the hands
7. Correction of anaemia
8. Treatment of osteoporosis
9. Protection of the fragile oesophagus, by treating gastric reflux
10. Prevention of constipation
11. Prevention of malnutrition with specialist dietetic advice required
12. Treatment of corneal and conjunctival blistering and ulceration
13. Specialist dental provision in view of oral blistering and ulceration with consequent microstomia
14. Regular skin surveillance with the aim of detecting squamous cell carcinomas.

Modes of inheritance and implications for obstetrics

Dominant Inheritance

With dominant transmission of a condition the patient themselves or their partner will be affected and there is a 1:2 chance with each pregnancy of the condition being passed on to any child the couple have together. There is no carrier status in a dominant condition and there is usually a long family history of EB. Occasionally an individual will present as the first person in his/her family to have the condition and they are said to have a de-novo mutation i.e. they have developed a new genetic mutation. (McClellan 2000)

Recessive Inheritance

With recessive transmission of a condition your patient or their partner will have inherited their condition from 2 unaffected parents. Their parents are both “healthy carriers” of a genetic mutation, which can cause EB. There is negligible risk (about 1:700) that your patients will have a child with EB. In order to do so their partner would have also to be carrying the mutation for EB, and the carrier rate is thought to be about 1:350 in the general population. However, it must be noted carefully that this picture can change if the pregnancy is the result of the relationship of close family e.g. 1st cousins or where the unaffected partner has a family history of EB. (McClellan 2000)

Pre-Natal Diagnosis

Pre-natal diagnosis for EB is usually offered to families who have had a child with a severe form of recessively inherited EB. This is because if one parent has a recessively inherited form of the condition there is no justification for offering pre-natal diagnosis as the likelihood of their children being born with EB is so small (see above).

In the dominant form of the condition pre-natal testing is only offered where a parent has a form of EB known as Dowling Meara EB Simplex. This is because infants with Dowling Meara EBS can be severely affected at birth. In other forms of dominantly inherited EB, pre-natal testing is not offered as on the whole it is felt that the conditions, whilst troublesome, are not so severe as to warrant potential termination of pregnancy.

Can the unaffected partner be tested for carrier status?

At present this is not a possibility, as a definitive answer cannot be given. This is because the mutations that lead to the recessive forms of EB are many. This is particularly so in the case of recessive dystrophic EB where the causative genetic mutation is found on the gene encoding for collagen VII. This is an enormous gene and the mutations leading to EB can be found anywhere on the gene. To look for an unknown mutation is akin to 'looking for a needle in a haystack' when you don't know what the needle looks like.

The fact that pre-natal and carrier testing is not available is frequently a source of great anguish for potential parents who have EB, particularly if they themselves are affected. The individual or couple will need careful counselling and should be referred to a specialist centre. (See Useful Contacts).

Care of the woman with EB.

The compromises and care needed by the woman with EB whilst she is pregnant and during childbirth will vary tremendously. The expert on your patient's condition is undoubtedly the patient herself and her opinion should be sought before carrying out any procedures. Other vital sources of information are the patient's Dermatology Consultant and the EB Nurse Specialists (see Useful Contacts). Whilst some women with EB have extreme skin fragility, others are less severely affected and only minor compromises are necessary when caring for them.

Care of the father with EB

Men with severe EB have become fathers. Our experience has been that fathers are concerned about the possibility of the baby being affected by EB and despite reassurance many of them have been plagued with anxiety about this throughout their partner's pregnancy. In dominantly inherited forms of EB it is clear that this is a very real anxiety. However if the baby does inherit EB it is almost always of a similar severity to the affected parent. Parents can be reassured that one form of EB cannot become another. In recessively inherited EB (as explained above) there is an almost negligible risk of the baby inheriting EB of 1:700. However for a father who has himself been affected by a very rare genetic disorder, it is understandable that his anxieties will be high. Fathers with EB may need encouragement to feel they will be able to participate in caring for their baby.

Early Pregnancy

Initially the major questions will be around modes of inheritance and issues about pre-natal diagnosis. It is important to ensure that the affected partner has had a firm diagnosis made at a specialist centre (see Useful Contacts). They should also have clear information on the mode of inheritance. If this is not the case they should be referred without delay.

The Antenatal Period

It is worth noting that some of the less desirable effects of pregnancy are also complications of some forms of EB. These include:

- Constipation
- Iron deficiency anaemia
- Oesophageal reflux.

For both comfort and well being these issues should be addressed and borne in mind when for instance giving analgesia.

Oesophageal Fragility

Morning sickness can be a real problem in some forms of EB, as can oesophageal reflux, because of the fragility of the oesophagus. Great care needs to be taken to prevent further damage and suitable medication prescribed. Many women with the severe forms of EB take proton pump inhibitors to control gastric acid, however these are not suitable for use in pregnancy.

Weight Gain

Many women with the severe forms of EB will be significantly underweight and will need specialist dietetic input particularly in pregnancy. This is as a result of a number of factors—increased nutrient requirement secondary to open wounds, and poor nutritional intake as a result of oral and oesophageal fragility. A Specialist EB Dietician is available at many of the EB centres

Blood Pressure

Please exercise caution when taking blood pressure as the cuff can damage skin. You may need to apply Melolin between the cuff and the skin.

Urinalysis

Occasionally blood may be found in urine due to open wounds in the perineal area or as a result of rare renal problems seen in EB (Atherton 2003) You should consult with the dermatologist involved if you suspect this problem is occurring for reasons other than obstetric ones.

Vaginal examinations

Proceed with great caution using well-lubricated gloves. This is particularly so if the woman has perineal involvement.

Abdominal examination

For some women with severe EB this may be a real problem as the pressure required in order to determine fundal height and discern foetal parts may blister their skin. This will be further exacerbated if you wear gloves and the woman's skin is moist. The application of cornflour (used in cooking) may aid this process. If there are open wounds please exercise great caution.

Wound infections

If the woman has a wound infection particularly in the abdominal area, if she is having LSCS, or in the groin area for a normal delivery, wound swabs should be taken and antibiotics may need to be given. Infections of wounds in the hands may put the infant at risk in the post-natal period and should be treated.

Help after delivery

Many families where one of the parents has a severe form of EB will need assistance in caring for their new baby. This is particularly so if there is hand involvement and where one parent is acting as carer for the parent with EB. Please speak to the DEBRA Social Care Team or EB Nurse Specialist if your patient feels they will need help in this regard. Planning in the early stages of the pregnancy is helpful as it often takes some time to secure funding and employ suitable carers.

Planning delivery

Some women with relatively severe forms of EB have had normal vaginal deliveries with few problems. However to date many women have had planned LSCS. Clearly this is a decision that has to be made by the women themselves, the obstetrician and midwife in conjunction with the EB team (Mallipeddi 2003).

Normal Delivery

- Carry out abdominal palpitation and vaginal examinations with great care. On carrying out vaginal examinations gloves should be well lubricated.
- Be cautious when attaching foetal monitoring equipment as tight belts may cause blistering.
- Do not use any adherent products
- When assisting your patient to move do not 'drag' them across sheets etc, as you will cause blistering/ remove skin
- Do not use incontinence pads as the plastic backing will contribute to skin blistering.
- If you need to catheterise the patient use a small well-lubricated catheter.
- Be aware that the perineal area may blister/shear away if it is handled less than gently.
- If using Entonox you may need to use Vaseline on the mouthpiece to prevent damage to the lips.
- If you are rubbing the patients back, do this with caution and apply powder or oil first to prevent skin damage.
- If you are holding the patients legs to assist her to deliver please powder your gloves or use a piece of soft gauze to protect the skin.
- Both episiotomies and 'tears' heal well in EB and their use is dictated by obstetric need.
- Breast-feeding may not be possible in the severer forms of EB if the nipples are very fragile. Some women do manage to breast feed despite their EB although this is the exception rather than the rule.

Skin and wound care

- No adhesive products of any kind must be used on the patients skin. If something is accidentally stuck to the patient or it is vital to remove an adherent product please remove using Apeel Spray or 50/50 or allow the product to fall off on it's own.
- Please ask the patient what they are able to tolerate. Specialist dressings will be supplied by the EB Nurse Specialist or the patient and include
 - > **Mepitel**—this can be cut into thin strips and used as tape to secure cannula etc along with a soft bandage. Mepitel is also used as a primary wound dressing and needs a secondary dressing. When handling Mepitel you are advised to moisten your gloves, otherwise the dressing will stick to you and not the patient.
 - > **Mepitac or Mepiform**—this can be used as tape and gives more secure fixation than Mepitel. It may therefore be suitable for securing epidural cannula. It would be cut into thin strips and removed with great care by rolling it back on itself rather than at right angles to the skin. However some patients feel this is too sticky for them and will not allow its use.
 - > **Mepilex**—this is a foam dressing which has some absorbency and may be suitable for dressing a LSCS wound.
 - > **Mepilex Lite**—this is thinner than Mepilex with slightly less absorbency.
 - > **Mepilex Transfer**—this is a very fine foam dressing which is suitable for low-exuding wounds only, unless it is used with a secondary dressing.
 - > All these dressings are made by Molnlycke and are coated with silicone to allow for ease of removal. All are now on prescription and most hospitals have at least some of them.

The patient will have a regular wound care routine and will have a supply of dressings at home. You may need to make arrangements for these dressing changes to be carried out if they are admitted to hospital. Some people with EB feel more secure if the dressing change is carried out by their usual carer and will be unwilling to let anyone else do the change. It may be advisable to discuss this early in the pregnancy in case of admission to hospital.

If anything does stick, do not panic and do not attempt to force removal. Does the adherent product have to be removed? - if not you can leave it to either fall off or allow the patients to remove it themselves. If you do need to remove it the application of a greasy substance such as 50/50 (white soft paraffin/liquid paraffin) will help.

Care during and following LSCS

1. If this is carried out under epidural anaesthesia then securing of the cannula should be as above.
2. If a general anaesthetic is given this must be carried out with great caution. The issues are avoidance of trauma to the mouth, oropharynx and larynx during intubation. The endotracheal tube should be well lubricated and Vaseline gauze should be used to protect the patients face/chin from the anaesthetists gloves/tubes etc. (Ames 1999)
3. EB patients are frequently difficult to cannulate, although the increased blood volume in pregnancy may go some way to ameliorating this problem. Secure IV lines with Mepitel and a soft bandage or Mepitac.
4. Unless vital do not apply ECG electrodes, pulse oximetry may be an option. If ECG electrodes are used it may be better to place them over the gel pads used under defibrillator paddles. If ECG electrodes are stuck directly onto the patients skin, **do not try to force removal**. Leave them to fall off or allow the patient to remove them themselves.
5. Only use airway suction if you have to, and try to use a soft catheter.
6. Do not stick diathermy pads to the patient, as alternatives are available.
7. Great care must be taken when moving EB patients, particularly if they are unconscious. They must be lifted or rolled gently, never dragged, as this will remove skin. Pat slides can be used with care.
8. LSCS wounds can be sutured or 'staples' as normal and should heal well. Please dress as above, and do not apply adhesive dressings.
9. Please be aware that your patient may need a pressure relieving mattress overlay particularly if they have had an LSCS.

Feeding

Most women with the severer forms of EB elect to bottle-feed because of the fragility of the skin of the nipples. However if your patient chooses to breast-feed she should be supported as we have had experience of women who have successfully breast-fed despite reasonably severe EB. This has however been a very difficult process with pain and blistering of the nipple. Correct 'fixation' of the baby to the breast is therefore extremely important in EB. Nipple shields may be of help in protecting the nipple.

Further information

DEBRA funds a team of EB Nurse Specialists who work with adults with EB. Please do contact the nurse working with your patient and she will be happy to discuss any aspect of your patients care. If your patient does not have a named nurse please contact the EB Nurse Consultant for your area (see useful contacts)

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