St John's Institute of Dermatology

Pregnancy and childbirth information for women with Epidermolysis Bullosa

Introduction

The care and management you will need whilst pregnant and during childbirth will vary enormously depending on what type of EB you have and how it affects you.

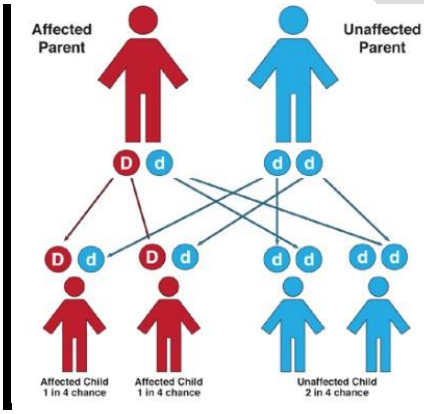
This information leaflet is intended to provide a general guide to some of the key things you are likely to encounter and questions you may have during your pregnancy. It supplements the professional guidance your obstetrician and midwives will require when managing your care.

Specialist advice and support both for you and for other healthcare professionals during your pregnancy is available from EB specialist teams so do make contact with your EB team if you are planning or have become pregnant.

Inheritance

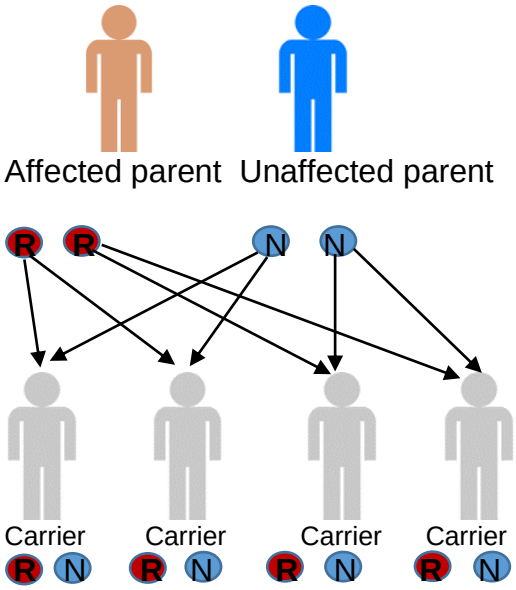
Whether you are likely to pass on your EB to your unborn child depends on the type of EB you have.

With dominantly inherited forms there is usually a long family history of EB and there is a 50% chance with **each** pregnancy of your child being affected. Typically EB Simplex (EBS) and Dominant Dystrophic EB (DDEB) fall into this category



Dominant inheritance pattern – one parent is affected with a dominant form of EB

If you have a recessively inherited type of EB (RDEB or Junctional EB) the risk of your child being affected is relatively small. In order for you to be affected with EB your parents were both healthy carriers of the recessive gene but were unaffected themselves. You will pass on one of the recessive EB genes to your baby, but it will only be affected by EB if your partner is also a **carrier** of the same faulty gene. **The carrier rate** is thought to be about 1:350 in the general population so the risk to your baby is small (Pillay 2006). In contrast the carrier rate of another well known recessively inherited condition, Cystic Fibrosis, is 1:25 (Cystic Fibrosis Trust 2017)



Inheritance pattern where one parent is affected by a recessive type of EB and one parent is unaffected and does not carry the recessive gene.
All children become carriers.

However if your partner is a close family relative (for example a first cousin) or if your partner's family also has a history of EB then the risk is considerably greater (McClean 2000). If this is the case then further discussion is recommended before you become pregnant

Pre-natal testing

This is not routinely offered however as advances are made in genetic screening, specialist centres may be able to offer this service.
Please talk to your EB nurse or doctor about the risk of your baby having EB and options for pre-natal testing.

Ante-natal diagnosis

Ante-natal testing is not routine but may be recommended to see if your baby has inherited EB. This is most likely to be advised if you have EB Simplex –Generalised Severe (formerly known as EBS Dowling Meara) or DDEB
Whatever type of EB you have, please talk to your EB team about the risk of your baby having EB and the options that are available to you.

Care during pregnancy

Morning Sickness can be a problem in early pregnancy for lots of women. In some types of EB fragility can extend internally and if this is a problem for you then frequent vomiting may cause blistering in your oesophagus. If you experience particular soreness after vomiting you should speak to your EB nurse for advice.

Weight gain is expected during pregnancy however some women, particularly those with the more severe forms of EB, may be underweight and will need specialist dietary advice and support during pregnancy. A specialist EB dietitian is available at many of the EB centres.

Ultrasound Scans are a routine part of ante-natal care. As long as you have intact skin over your abdomen there should have no problem with standard ultrasound scans. Just remind the scanner to take care when wiping off the gel so that they don't rub too much.
If you have a more severe form of EB, scanning may be more difficult as the pressure required on your abdomen to see the foetus may cause blistering to your skin. If you are worried about having a scan get in touch with your EB nurse who will be able to support you and offer advice to the person taking the scan.

Urinalysis. You will be asked to bring a urine sample to every maternity appointment. If you have any open wounds in the perineal area then the midwife may detect blood in your urine. There can also be pregnancy linked reasons for the blood which should be investigated, but you should also alert your midwife to the fact that it may be related to your EB.

Vaginal examinations can be carried out but you need to ask the midwife to be very careful, particularly if you have any perineal involvement. Make sure the midwife uses well-lubricated gloves.

Planning your delivery

Whether to have a normal vaginal delivery or caesarean section should depend on you and how your pregnancy has progressed rather than on the type of EB you have (Choi 2011). The decision about whether to have a normal delivery or to opt for a caesarean section should be taken after talking with your obstetrician, midwife and the EB team. Some women with relatively severe EB have had normal vaginal deliveries with few problems, however many women opt to have a planned caesarean section (Choi 2011).

An EB pregnancy information pack can be provided by your EB nurse; this will include relevant professional guidelines for your obstetrician and midwife, contact details for your EB nurse and some useful dressings. You should keep this with your "labour bag" and make sure you share it with whoever is looking after you during your delivery.

Normal vaginal delivery

You have the best understanding of your EB and what you are able to tolerate. Your midwife's priority will be to provide the best care for you and ensure the wellbeing of your baby, but they may have only very limited understanding of EB. Being well prepared ahead of labour will be invaluable but it can also be useful if you make sure your birth partner also has a clear understanding of how to manage delivery with EB in mind as they may need to speak up and be your advocate.
Make sure you share the professional guidelines from your EB pregnancy pack with your midwife. If you have one of the more severe forms of EB then discussions and planning between your obstetrician, midwife and your EB nurse well in advance of your delivery would be extremely valuable. Your EB nurse will be able to help to coordinate this.

Caesarean section

Following consultation with your obstetrician, midwife and your EB team you may make the decision to have a caesarean section.

With most types of EB managing a caesarean section will be much the same as for any surgical intervention and should be straightforward. Ideally the section will be planned, but it is always worth thinking ahead about what to do if you need an emergency procedure.
Surgical wounds in all types of EB generally heal well and sutures or clips can be used to close your caesarean wound in exactly the same way as for women who do not have EB.
If you have a more severe form of EB your pregnancy and delivery will need to be much more closely monitored and supervised. Since pregnancy in women with Recessive Dystrophic Epidermolysis Bullosa is rare, information about the best way to deliver your baby is limited; however what is absolutely essential is thorough assessment, preparation and coordination of your care (Turmo-Tejera 2014) and your EB team should support you throughout your pregnancy to make sure this happens.

Feeding

Most women with more severe forms of EB choose to bottle feed because of the fragility of the nipple area. If you do choose to breast feed your midwife will need to work closely with you to help your baby to latch on properly and you should expect pain and blistering of the nipple. Some women have found nipple shields to be helpful.

The new baby with EB

If you suspect your baby has inherited EB then you should contact the paediatric EB team at either Great Ormond Street Hospital (0207 829 7808) or Birmingham Children's Hospital (0121 333 8224).
Guidance about the care of a new baby with EB can be found at:
http://www.gosh.nhs.uk/healthprofessionals/clinical-guidelines/epidermolysis-bullosa

Support and further information

DEBRA
Debra House, 13 Wellington Business Park, Dukes Ride, Crowthorne, Berks RG45 6LS
01344 771961

Adult Nursing Teams
St Thomas' Hospital, Lambeth Palace Road, London SE1 7EH
EB Administrator 0207 188 0843

Heartlands Hospital, Lode Lane, Solihull B91 2JL
0121 424 2000

Useful guidelines for health care professionals
Surgical guidelines – useful in event of a Caesarean Section
http://www.debra.org.cn/fileadmin/user_upload/medresearch/2016/surgical-procedures.pdf
Best Practice in care of patients with EB. Covers complex management of EB and includes lots of information about suitable dressings
http://www.woundsinternational.com/best-practices/view/best-practice-guidelines-skin-and-wound-care-in-epidermolysis-bullosa

References

Choi, S.D., Kho, Y.C., Rhodes, L.M., Davis, G.K., Chapman, M.G. and Murrell, D.F. (2011) Outcomes of 11 pregnancies in three patients with recessive forms of epidermolysis bullosa. *British Journal of Dermatology,* 165: 700–701.

Cystic Fibrosis Trust www.cysticfibrosistrust.org.uk (accessed September 2017)

McClean I (2000) *The genetics of EB.* Debra UK.

Pillay E (2006) *Care of the woman with EB during pregnancy and childbirth*. Debra UK.

Turmo-Tejera, Marta; García-Navia, Jusset T; Suárez, Francisco; Echevarría-Moreno, Mercedes. (2014) Cesarean delivery in a pregnant woman with mutilating recessive dystrophic epidermolysis bullosa. *Journal of Clinical Anesthesia;* Philadelphia (26.2) , 155-157

The Epidermolysis Bullosa service runs in collaboration with the charity DEBRA

DEBRA is the national charity supporting those directly affected by, and working with, Epidermolysis Bullosa (EB) – a potentially fatal skin condition that causes constant pain due to unstoppable internal and external blistering. A charity registered in England and Wales (1084958) and Scotland (SC039654). Company limited by guarantee registered in England and Wales (4118259).

Contact us

Adult EB Clinical Nurse Specialist team
St Thomas' Hospital, Lambeth Palace Road, London SE1 7EH
Contact via EB Administrator on 0207 188 0843

Monday to Friday 8am -6pm

http://www.guysandstthomas.nhs.uk/our-services/dermatology/specialties/adult-epidermolysis-bullosa/overview.aspx

Pharmacy Medicines Helpline
If you have any questions or concerns about your medicines, please speak to the staff caring for you or call our helpline.
t: 020 7188 8748 9am to 5pm, Monday to Friday

Your comments and concerns
For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.
t: 020 7188 8801 (PALS) **e:** pals@gstt.nhs.uk
t: 020 7188 3514 (complaints) **e:** complaints2@gstt.nhs.uk

Language and accessible support services
If you need an interpreter or information about your care in a different language or format, please get in touch:
t: 020 7188 8815 **e:** languagesupport@gstt.nhs.uk

NHS 111
Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day.
t: 111

NHS Choices
Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.
w: www.nhs.uk

Get involved and have your say: become a member of the Trust
Members of Guy's and St Thomas' NHS Foundation Trust contribute to the organisation on a voluntary basis. We count on them for feedback, local knowledge and support. Membership is free and it is up to you how much you get involved. To find out more, and to become a member:
t: 0800 731 0319 **e:** members@gstt.nhs.uk **w:** www.guysandstthomas.nhs.uk/membership

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