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Retrospective evidence on outcomes and experiences of pregnancy and childbirth in epidermolysis bullosa in Australia and New Zealand☆,☆☆,★

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ABSTRACT

Background: Pregnancy in epidermolysis bullosa (EB) has not been comprehensively studied.
Objective: We aimed to develop a foundational database, which could provide peri-obstetric advice in EB.
Methods: Survey questionnaires were sent to obstetricians, unaffected mothers of EB babies, and mothers with EB. Results were analyzed using chi-square, Fisher exact, and t-tests.
Results: Out of 1346 obstetricians surveyed, 195 responded, and only 14 had encountered EB. All recommended normal vaginal delivery (NVD), except for one elective Caesarean section (CS). We received responses from 75 unaffected mothers who had delivered EB babies. They had significantly more complications in their EB pregnancies compared to their non-EB pregnancies. A further 44 women with various types of EB who had given birth responded. Most delivered via NVD and had no significant increase in complications in both their EB and non-EB pregnancies. In both groups, there were no significant differences in blistering at birth in babies delivered via NVD and CS.
Conclusion: In conclusion, most patients with EB who are capable of giving birth do not have an increased risk for pregnancy-related complications and NVD appears to be safe. Awareness of this data amongst obstetricians and dermatologists should lead to improved quality of care for mothers and babies affected with EB.

Introduction

Ongoing research on various aspects of epidermolysis bullosa (EB) is currently underway. Most reports are focused on the molecular basis and classification of this disease. Diagnostic criteria and treatment options for this condition are constantly evolving, but little focus has been directed towards pregnancy and childbirth in these patients. There is a scarcity of literature available addressing this important issue, and this paper aims to fill that gap, and provide sound evidence and guidance for mothers who are pregnant with EB babies.

Milder forms of EB, such as EB simplex, often go undiagnosed: this could explain the relative lack of pregnancy cases reported. On the other hand, very few reports in the literature detail pregnancy and childbirth experiences of mothers and infants with more severe forms of EB, including junctional EB (JEB) and recessive dystrophic EB (RDEB). It does not always follow that all patients with severe forms of EB will have difficult pregnancies (Price and Katz, 1988).

The bulk of the available literature is mainly on prenatal diagnosis of severe forms of EB (EBS or JEB with pyloric atresia, Herlitz JEB, and RDEB) and its role in management decisions such as termination (D'Alessio et al., 2008; Marinkovich et al., 1995; Yan et al., 2007; Pfendner et al., 2003; Norup 1999). A survey performed in Denmark amongst obstetricians and pediatricians showed that in the case of newborns with severe EB, there was a strong consensus to withhold life-prolonging treatment, reflecting attitudes to EB (Norup 1999).

A patient with non-Herlitz JEB was reported who had two miscarriages prior to giving birth successfully via Cesarean section under epidural anesthesia (Price and Katz, 1988). A patient with RDEB in Germany had two vaginal deliveries resulting in healthy babies, with uncomplicated episiotomy wound healing, and no exacerbations of EB during her

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pregnancy (Büscher et al., 1997). Another patient with RDEB had preterm labor at 36 weeks and premature rupture of membranes, yet delivered a healthy baby via Cesarean section (Bianca et al., 2003). In the French literature, there is a report of a patient with EBS who developed a herpetiform flare of EBS-DM during the first two months of her pregnancy (Diris et al., 2003). More recent reports include that of a patient with Kindler syndrome with vaginal stenosis who had a successful Cesarean delivery (Hayashi et al., 2007). The report most recently published is a case report of 11 pregnancies in three patients with recessive EB in Australia. One of the patients had non-Herlitz JEB and had delivered two unaffected babies via NVD eight years apart. The two other patients were sisters who both had generalized RDEB. One of them delivered three healthy unaffected babies via NVD, and the other delivered five unaffected babies via NVD. They all had no complications or flare of their EB during their pregnancies and the peripartum period (Choi et al., 2011). More recently, there has been a report of three more women, each with RDEB-intermediate (RDEB-I), all of whom had successful vaginal deliveries without major cutaneous or mucosal complications (Hanafusa et al., 2012). There is also an online patient information handout on pregnancy and childbirth in EB published by the Dystrophic EB Research Association (DEBRA) UK group in May 2006 which reports that women with EB have successfully had vaginal and Cesarean deliveries (Pillay, 2006).

Labor and delivery practices include airway management strategies, the role of regional anesthesia, and the use of nonadhesive tape and padding (i.e. Mepitel, Mepilex, Mepitac, Mepiform) as minor trauma may lead to severe lesions (Price and Katz, 1988; Pillay, 2006). Regional anesthesia has been used successfully in these patients. There are five reported cases that used either spinal or epidural anesthesia for Cesarean section, and epidural anesthesia for vaginal delivery without any ensuing complications (Baloch et al., 2008; Broster et al., 1987; Berryhill et al., 1978).

In view of this limited information, we designed a survey looking at the experiences of a large group of obstetricians, unaffected mothers who delivered babies with EB, and EB patients themselves who have delivered babies. We have developed a foundational database, and have developed recommendations on periobstetric advice in relation to EB.

Methods

This study was granted ethics approval by the South Eastern Sydney Local Health District Human Research Ethics Committee - Southern Sector on the 3rd of October, 2006 until July, 2012.

Questionnaires were sent out to three participant groups, namely obstetricians in Australia, unaffected mothers who had given birth to EB babies, and EB females who had given birth.

The list of obstetricians was obtained from the Royal Australian and New Zealand College of Obstetricians and Gynecologists (RANZCOG), whilst the list of EB patients and their parents was obtained from patients known to us, most of whom are in the Australasian EB Registry which is being maintained at St. George Hospital, Sydney, NSW, Australia (Kho et al., 2010). The appropriate questionnaires were mailed to these obstetricians and patients in the post with self-addressed envelopes. Some questionnaires were also handed out to member families of DEBRA Australia and New Zealand, new patient referrals seen at St. George Hospital, and patients attending EB clinics. All participants had given signed informed consents to participate in the study and share their data.

A substudy was also performed that looked into the percentage and locations of blisters, if any, in babies born with EB to unaffected mothers and those diagnosed with EB This was achieved by sending out further questionnaires with body maps to both groups of respondents.

The data was then collated and summarized over a period of 4.8 years (October 2006–August 2011). Statistical analysis was performed using chi-square tests, t-tests, and Fisher exact tests. The statistical program used was SigmaStat. Based on the results, peri-obstetric recommendations were made for EB patients and mothers giving birth to EB babies.

Results

Group 1: Data from obstetricians in Australia

The questionnaires were sent out in one batch to 1346 obstetricians in Australia, and 195 responded. Only 14 of the 195 obstetricians who responded had encountered mothers or babies born with EB. Their average number of years in practice was 17. Six of the 14 obstetricians attempted a literature search on EB in pregnancy and childbirth, but only three were successful in finding any articles on pregnancy in EB. Also, only four had coordinated the management of these patients with a dermatologist.

The 14 obstetricians all recommended normal vaginal delivery (NVD). However, one performed an elective Cesarean section (CS) at the patient's request which resulted in poor wound healing and a post-operative wound infection. Furthermore, 111 of the 195 obstetricians indicated the need to have information about EB available in antenatal clinics.

Group 2: Data from mothers without EB who had given birth to EB babies

We sent 122 survey questionnaires to EB-unaffected mothers who had given birth to at least one child with EB. Attempts were made to contact all non-responders, and they were re-sent the survey forms. We received 75 completed questionnaires out of the 110 mailed out (a 68% response rate or 75% response of those known to us). An additional 12 forms were returned, undelivered, owing to changes of address. These pertained to 176 pregnancies and 174 births, 84 (48%) of whom were affected by various types of EB: (35 with EBS, 14 with JEB, 19 with DDEB, and 17 with RDEB). There were 69 surveys from mothers in Australia and 6 from New Zealand. The age of respondents ranged from 19–78 (mean age of 46.2) and they had given birth to between one and four children, with an average of about two children in each family. A total of 43 out of the 75 mothers (57%) had children under 18 years of age. A summary of their characteristics is shown in Table I.

Most mothers with EB had normal vaginal deliveries without any ensuing complications. The ratio of NVD to CS was 4:1. Table II shows the modes of delivery of both EB-affected and EB-unaffected children whilst Table III shows the list of complications during pregnancy and delivery of both groups of babies from mothers who were unaffected by EB themselves. A chi-square analysis showed significantly more (approximately two-fold) complications in the pregnancies that delivered EB babies (22/84) as compared to EB-unaffected babies (11/90) ($p = .031$). Fisher's exact testing also revealed significantly more complications in EB babies delivered via CS (including emergency CS) as compared to NVD ($p < .001$).

Five cases of full-term babies not known to have EB in advance due to a lack of family history of EB were delivered using vacuum suction and/or forceps, resulting in skin being eroded from the babies' head, face and mouth areas; these babies were subsequently diagnosed with severe forms of EB. The first case was a baby with RDEB delivered via NVD and vacuum suction in which skin was removed from the baby's face and mouth. The second case was a baby with RDEB delivered via NVD and forceps, where skin was removed from non-facial parts of

Table I
Demographics of mothers with EB, and mothers without EB, who have given birth to EB babies.

	Number of mothers with EB	Number of mothers without EB
Number of mothers	44	75
Mean age and age range	45.1 (22-82)	46.2 (19-78)
Mothers from Australia	37	69
Mothers from New Zealand	7	6
Number of offspring	112	174
EB-affected babies	54	84
EB-unaffected babies	58	90

EB, epidermolysis bullosa.

Table II
Modes of delivery in unaffected mothers of EB babies.

Mode of Delivery	Babies born with EB					Unaffected
	EBS	JEB	DDEB	RDEB	Total EB[1]	
NVD	26	11	17	13	67 (80%)	73 (81%)
PCS	7	0	1	2 (12%)	10 (12%)	10 (11%)
ECS	2 (6%)	3 (21%)	1(5%)	1 (6%)	7 (8%)	7 (8%)
Total deliveries	35	14	19	16	84	90

EB, epidermolysis bullosa; NVD, normal vaginal delivery; PCS, Planned Cesarean Section; ECS, emergency Cesarean section.
[1] Percentages of actual deliveries in parentheses.

the baby's head. The third case was a baby with JEB delivered via NVD and forceps, which resulted in skin erosion from the baby's face. The fourth case was a baby with JEB delivered via NVD and vacuum suction resulting in skin being eroded from the baby's feet. The fifth case was a baby with JEB delivered via emergency CS using both vacuum suction and forceps, resulting in facial erosions and hematomas. The overall rate of emergency CS was 8% but for pregnancies with JEB offspring it was 21%, suggesting that labor in this group is more complicated. Out of the 84 babies with EB, 46 (55%) had blisters at birth, most commonly in the severe types of EB (JEB and RDEB) and the others (45%) developed blistering in the days or weeks subsequently.

Most mothers surveyed were unaware that they were going to deliver a baby with EB. Some expressed the view that their obstetrician could have given more accurate information regarding the genetics and severity of the EB type that affected their babies. Other mothers, particularly with children affected by severe EB types recommended prenatal screening and informed decisions about termination options. Those whose babies had blistering due to birth trauma thought to be associated with vaginal delivery recommended delivery via CS.

Group 3: Mothers with EB who had given birth

Out of 55 females with EB of childbearing age surveyed, 44/55 (80%) returned completed questionnaires. We tried to contact all 11 non-responders, and sent out new surveys forms. Three were returned unanswered due to change of addresses. The revised response rate of 44/52 or 85% of contactable female EB patients of childbearing age was very high. For those known to have at least one child also affected with EB (32/36), the response rate was 89%. We received responses from 28 patients with EBS, 1 with JEB, 12 with DDEB and 3 with RDEB. They had given birth to a total of 112 babies, 54 affected with dominant types of EB (37 with EBS and 17 with DDEB) and to 58 babies unaffected by EB. The average age of respondents was 45.1 years (range, 22–82 years). The respondents' birth rates averaged about two per family, varying between one and eight children. The proportion of mothers with children currently under 18 was 24/44 (55%). A summary of their characteristics is shown in Table I.

In general, the mothers' EB conditions remained stable during pregnancy and the immediate peripartum period. Only two of the 44 patients, both with EBS, reported that it worsened. Two others (1 with EBS and 1 with RDEB) reported improvement in their condition.

Most patients had normal vaginal deliveries with minimal complications. The ratio of NVD to CS was 4:1. Table IV compares the modes of delivery in this group of patients, and Table V lists complications. There were fewer complications during their pregnancies with EB-affected babies (8/54), compared to their pregnancies with unaffected babies (15/58). However, a chi-square test showed no significant difference in the number of complications for these two groups ($p = .225$).

EB patients who delivered via NVD (91/112 deliveries, 81%) all reported good healing of their episiotomy incisions and perineal tears, where occurring. During delivery, four EBS patients reported blistering at sites where adhesive tape was used to secure their epidural anesthesia. Post-delivery, 10 patients (8 with EBS, 1 with JEB, and 1 with RDEB) reported nipple blistering while breastfeeding, which led them to switch to bottle-feeding. The EB mothers who delivered via CS (21/112 or 19%), had good healing of their CS incision sites with just two reports of post-operative wound infections, which later healed well.

Table IV
Modes of delivery of mothers with EB who had given birth.

Mother's Type of EB*	Mode of Delivery		
	NVD	PCS	ECS
EBS (28)	**55**	**6**	**10**
Infant with EBS	*33*	*1*	*3*
Infant without EBS	*22*	*5*	*7*
JEB (1)**	**2**	**0**	**0**
DDEB (12)	**26**	**0**	**4**
Infant with DDEB	*15*	*0*	*2*
Infant without DDEB	*11*	*0*	*2*
RDEB (3)**	**8**	**1**	**0**
Total	**91**	**7**	**14**

EB, epidermolysis bullosa; NVD, normal vaginal delivery; PCS, planned Cesarean section; ECS, emergency Cesarean section; JEB, junctional epidermolysis bullosa; RDEB, recessive dystrophic epidermolysis bullosa.
* in parentheses is the number of mothers with EB; *italics* = their offspring.
** all offspring of JEB and RDEB females unaffected.

Table III
Complications during pregnancy and delivery of EB babies by mothers without EB.

Complications	Unaffected babies ($n = 90$)	NVD	PCS	ECS	EB Babies ($n = 84$)	NVD	PCS	ECS
Bleeding	0	0	0	0	1	1	0	0
Cord coil around neck	0	0	0	0	1	1	0	0
Emergency CS for cephalopelvic disproportion	1	0	0	1	0	0	0	0
Emergency CS for other reasons	4	0	0	4	2	0	0	2
Gestational diabetes	1	1	0	0	4	3	1	0
Hyperemesis	0	0	0	0	3	2	0	1
Hypertension	1	0	1	0	0	0	0	0
Hypoglycaemia	1	1	0	0	0	0	0	0
IUGR	0	0	0	0	2	1	1	0
Oligohydramnios	0	0	0	0	1	1	0	0
Placenta previa	0	0	0	0	2	0	0	2
Polyhydramnios	1	0	0	1	0	0	0	0
Preeclampsia	1	0	1	0	3	0	1	2
Preterm labour	1	1	0	0	2	1	1	0
PUPPP[2]	0	0	0	0	1	1	0	0
Total	11 (12%)[1]				22 (26%)[1]			

EB, epidermolysis bullosa; PCS, Planned Cesarean Section; ECS, Elective Cesarean Section.; IUGR, Intrauterine growth retardation; PUPPP, pruritic urticarial papules and plaques of pregnancy.
[1] $p = .03$

Table V
Complications during pregnancy and delivery of EB babies by mothers with EB.

Complication	Unaffected babies ($n = 58$)	NVD	PCS	ECS	EB babies ($n = 54$)	NVD	PCS	ECS
Abruptio placenta	1	1	0	0	0	0	0	0
Bleeding	2	1	1	0	1	1	0	0
Cholestasis	1	1	0	0	0	0	0	0
Emergency CS for cephalopelvic disproportion	1	0	0	1	1	0	0	1
Emergency CS for other reasons	3	0	0	3	2	0	0	2
Fetal distress	0	0	0	0	1	1	0	0
Gestational diabetes	1	0	1	0	0	0	0	0
Hypertension	1	1	0	0	0	0	0	0
Placenta previa	0	0	0	0	2	2	0	0
Preeclampsia	2	2	0	0	1	0	0	1
Preterm labor	1	0	1	0	0	0	0	0
Prolonged labor	2	1	0	1	0	0	0	0
TOTAL	**15 (26%)***				**8 (15%)***			

ED, epidermolysis bullosa; PCS, planned Caesarean section; ECS, elective Caesarean section; CS, Caesarean section.
* $p = .225$

General advice from mothers with EB was to have genetic testing done in case a more serious type could be passed on. However, the general consensus was that the joy of having children was worth the discomfort and pain.

Sub-study of Blistering at Birth from Groups 2 and 3

We sent out further questionnaires to the participants in Group 3 (EB females who had given birth) to look at blistering at birth in EB-affected babies in relation to their mode of delivery. The questionnaire included body maps. The respondents were asked to shade areas affected by blistering. A total of 32/44 Group 3 mothers had given birth to babies affected with dominant forms of EB, and from this group, we received 13/32 responses. This subgroup of respondents from Group 3 had given birth to 19 babies with EB (14 with EBS and 5 with DDEB) and 14 unaffected babies. The mode of delivery was mostly NVD.

Table VI shows the number of babies born with blisters at birth in relation to their mode of delivery. In this table, data were combined from Groups 2 and 3. Fifty-five percent (46/84) of EB babies born to EB-unaffected mothers (Group 2) had blisters at birth. Similarly, 58% (11/19) of EB babies born to mothers with EB (Group 3) had blisters at birth. Fisher's exact test showed that blistering in JEB and RDEB babies is significantly higher than in EBS babies ($p = .016$ and $p < .001$ respectively). Blistering in DDEB babies was not significantly greater than blistering in babies with EBS ($p = .769$). Fisher's exact test showed blistering at birth in RDEB babies is significantly more common than in DDEB babies ($p = .012$). Finally, blistering in JEB babies was not significantly different from blistering in RDEB babies ($p = .315$). In addition, there were no significant differences ($p = .121$) in blistering of RDEB babies delivered via NVD and planned CS. Overall, there was no significant difference in blistering at birth in all EB babies delivered via NVD versus CS ($p = .136$).

Table VI
Proportions of EB babies born with blisters at birth in relation to their mode of delivery.

Type of EB	Blisters at birth	NVD	PCS	ECS
EBS	20/49 (41%)	14/37 (38%)	4/7 (57%)	2/5 (40%)
JEB	11/14 (79%)	8/11 (73%)	0/0 (0%)	3/3 (100%)
DDEB	11/24 (46%)	9/21 (43%)	0/1 (0%)	2/2 (100%)
RDEB	15/16 (94%)	12/13 (92%)	2/2 (100%)	1/1(100%)
Total	**57/103 (55%)**	**43/82 (52%)**	**6/10 (60%)**	**8/11 (73%)**

EB, epidermolysis bullosa; EBS, epidermolysis bullosa simplex; JEB, junctional epidermolysis bullosa; DDEB, dominant dystrophic epidermolysis bullosa; RDEB, recessive dystrophic epidermolysis bullosa; PCS, planned Caesarean section; ECS, emergency Caesarean section; NVD, normal vaginal delivery; CS, Caesarean section.
Modified to include responses of Unaffected Mothers of EB babies ($n = 75$) and Females with EB who have given birth to babies with EB ($n = 13$).
No significant differences in blistering at birth in all EB babies delivered via NVD versus CS, $p = .136$.

Discussion

An international expert consensus on delivery recommendations for patients with EB or for EB-unaffected mothers expecting infants with EB has yet to be established. Hence, this survey is quite timely. Due to the rarity of this family of diseases, there was a relatively low response rate amongst obstetricians, most of whom felt that NVD should be the recommended mode of delivery for EB patients giving birth. Data from a larger prospective cohort study within the 2005 WHO global survey on maternal and perinatal health have shown that, overall, maternal morbidity and mortality were higher in the elective CS group (5.5%) than the NVD group (1.8%). Furthermore, increased risk in NVD relates to maternal socio-demographic characteristics such as being single, young with a low level of education, gravidity, and primiparity. Increased risk for maternal morbidity and mortality in the CS group related to women with previous complications in their pregnancies or perinatal outcomes (Villar et al., 2007). This supports our data that NVD is still the recommended mode of delivery for most mothers carrying EB babies and for pregnant EB females. Despite this recommendation, there seems to be a growing preference for elective delivery by CS, particularly in Western countries.

In a recent structured survey performed to determine personal preferences of delivery method amongst obstetricians from Australia and New Zealand (which had a 26% response rate), 11% of obstetricians chose elective CS in the absence of any clinical indication. Elective CS procedures were also the preferred method of childbirth in cases of predicted fecal incontinence (83.5%), urinary incontinence (81.5%), perineal damage (68.5%), and fear of damage to the baby (24%) (Land et al., 2001).

The rates of CS in most developed countries are quite similar, with 23.3% of all births in Australia, 21.3% in the UK and 26% in the US (Dodd et al., 2007) performed by CS. Overall, NVD is still the most recommended mode of delivery worldwide and appears to be the safer method of childbirth. It should be emphasized, however, that forceps delivery or vacuum suction should be avoided during NVD or CS, as our data have shown that babies with severe forms of EB had severe erosions on their head and feet. The data also suggest that Cesarean wounds heal well in mothers with EB, and that care during breastfeeding (i.e. use of nipple shield) or bottle-feeding, are recommended options if blistering is severe.

As for applicability of data derived from the mothers of children with EB, our response rate of 75% from mothers of children with EB is significant, given that the average response rate cited in the literature for mailed physician questionnaires is around 61% and this has remained quite stable over time (Cummings et al., 2001). Hence, the results of our data collection should have excellent applicability. Interestingly, the surveys of mothers who gave birth to babies with EB reveals that there were significantly more complications in deliveries by CS

compared to the majority who delivered via NVD. This was particularly true for infants with JEB or RDEB. Overall, either of the two modes of delivery seemed to be comparable for blistering rates. Blistering at birth in the different types of EB showed the more severe forms of EB (JEB and RDEB) had significantly more blistering than the milder EBS and DDEB forms, as might be expected. Together, this suggests that if it were known in advance that a mother was pregnant with a baby with EB, delivery via NVD would still be recommended as the preferred mode of delivery as long as it is safe to do so; for example, providing that cephalopelvic disproportion is not a problem. This would be the case for 50% of mothers with a dominant form of EB such as EBS and DDEB. It would be more difficult, if not impossible, to predict complications in those with no known family history of a recessive form of EB. Genetic counseling and discussion of prenatal diagnostic options are recommended for all EB patients when contemplating pregnancy (Sybert, 2010; Fassihi and McGrath, 2010).

Conclusions

Most patients with EB are capable of giving birth without increased risk of pregnancy-related complications. Unaffected mothers who have given birth to children with EB have had relatively normal pregnancies comparable to previous pregnancies yielding unaffected children. However, when a mother is known to be carrying an EB pregnancy, delivery via normal vaginal delivery is no more likely to result in complications and blisters at birth in the EB-affected newborn. Hence, there appears to be no justification in performing a Cesarean section to reduce complications for the mother with EB nor the infant with EB in order to avoid EB-related complications. CS should be reserved for the traditional indications of all pregnant mothers. Lastly, awareness of these data amongst obstetricians and dermatologists should lead to informed advice and improved quality of care for both EB mothers and EB babies alike.

Recommendations for expectant patients with EB

1. Normal vaginal delivery with regional anesthesia is generally safe, and episiotomy may reduce perineal tears.
2. Vacuum suction or forceps delivery is not recommended in mothers delivering babies with EB or where the EB status of the baby is unknown.
3. In mothers expecting to deliver a baby with EB, normal vaginal delivery is still the preferred mode of delivery.
4. Only non-adhesive tape and dressings are to be used during anesthesia and surgery.

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