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Assessment of Epidermolysis Bullosa Awareness and Knowledge in Hail City Population in Saudi Arabia

Fawwaz F. Alshammarie[1], Turki S. Aljuhani[2], Abdulrahman F. Alluhaybi[3]

[1]Assistant Professor, Department of Dermatology, College of Medicine, University of Hail, Hail, Saudi Arabia.
[2]Dermatology Residents, Ad Diriyah Hospital, Riyadh, Saudi Arabia.
[3]Dermatology Residents, King Saud University Medical City, Riyadh, Saudi Arabia.

ABSTRACT

Background: *Epidermolysis Bullosa is inherited disease that has different subtypes, some of which have catastrophic outcomes. Incidence is increasing in Hail city. Hence, our aim is to assess the population's awareness of this disease.*

Methods: *Over the course of five months, this study was conducted among Hail's population. A detailed questionnaire, concentrating on general viewpoints of Epidermolysis Bullosa, was made, and piloted before the final interview. A total of 517 resident were involved in the study.*

Results: *Out of 517, 56% were males and 43% females. There were 27.3% of respondents between 32-25 years of age, followed by 23-24 years of age (26.3%), 33-40 years (20.7%), elderly (14.1%), with only 11.6% of respondents under the age of 18. Most participants (89%) have a low level of knowledge about Epidermolysis Bullosa, according to the results.*

Conclusion: *Our study showed that Hail city population had poor level of knowledge about EB. Through educational campaigns or any other educational method that is possible, the Hail city population and caregivers in particular need to be aware of EB and its consequences.*

KEYWORDS: *Epidermolysis Bullosa, Population, Knowledge, Awareness, Hail, Saudi Arabia*

INTRODUCTION

Inherited Epidermolysis Bullosa (EB) includes many genotypically different entities that share a common characteristic feature which is fragility of epithelial lined tissues due to minor trauma, particularly in the skin (Lanschutzer, 2008). Many cutaneous findings that may be seen in EB include: milia, alopecia, dystrophy or nonappearance of nails, exuberant granulation tissue, aplasia cutis congenita, mottled pigmentation, palmoplantar keratoderma, and EB nevi (Lanschutzer, 2008). EB simplex, junctional EB, dystrophic EB, and Kindler syndrome are the four most common types of inherited EB. (Jo David Fine et al., 2008). Hail Patinets primarily had dystrophic EB and there were two linked homozygous mutations also found in the same gene: G1400Vfs*310 and c.1370C>T, P457L. (Alshammari et al., 2019). In general, routine light microscopy is of slight value in the diagnosis of hereditary EB and diagnostic immunofluorescence antigenic mapping and transmission electron microscopy are preferred methods of diagnosis (Lanschutzer, 2008). The prevalence and incidence of EB in the United States of America approximately 8.2 per 1 million and 19.6 per 1 million live births , respectively (Fine, 2016).

In Saudi Arabia, the prevalence and incidence of the disease are unknown. Meanwhile, we are seeing new cases in Hail city. One of the major causes is the consanguineous marriage. Therefore, we should raise the awareness among the population. The major underlying principles of care for all EB patients are avoidance of blistering by cautious protective padding of the skin and prevention of secondary infection as much as possible by careful wound care, using sterile synthetic non-adhesive hydrocolloid bandages (Lanschutzer, 2008).

METHODS AND MATERIALS

The present study is a quasi-experimental, cross-sectional, questionnaire-based study, conducted over five months (May to October 2020) among dwelling population of Hail, Saudi Arabia. A detailed questionnaire containing demographic variables and concentrating on assessing the awareness and knowledge of participants regarding Epidermolysis Bullosa, was made, and piloted before the final interview. The study involved 517 residents. The questionnaire section assessing the knowledge of participants regarding EB consisted of 11 questions. One point was given for every correct answer and zero for incorrect answers or the option "I do not know." for

each correct answer given by the participant, one point was given to him, and pointes were added up for a maximum of 11 points. Therefore, the level of knowledge was classified as poor (0-4), moderate (5-8), and good (9-11). The study was conducted after obtaining the ethical approval from the Ethics committee, college of medicine, University of Hail, KSA. The participation in this study was voluntary and the questionnaires were kept anonymous. Before participation, the aim of this study was explained to participant. All the statistical analysis was carried out on SPSS 16.0 version (chicago, inc., usa), where frequencies and percentages were conducted to illustrate qualitative and categorical variables, while chi-square test was used for comparisons between dependent and independent variables.
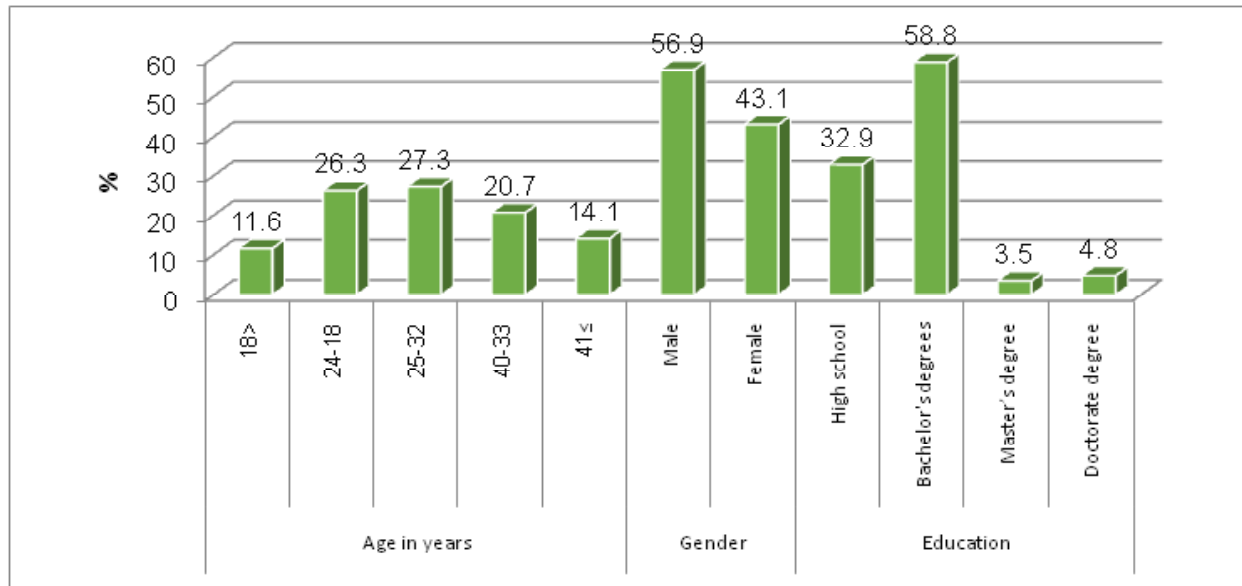
RESULTS

Figure. 1. distribution of respondents according to age, gender and educational level

As shown in Figure 1, The total number of participants were 517 including 294 (56.9%) males and 223 (43.1%) females. Most of respondents were between the age of 32-25 years (27.3%) followed by the age of 18-24 (26.3%), 33-40 (20.7%), age of more than 41 year (14.1%), while only 11.6% of respondents below the age of 18 year.

Table 1. distribution of respondents' awareness about etiology of epidermolysis bullosa disease and its risk factors

Have you ever heard of epidermolysis bullosa?	No. (n=517)	%
Yes	58	11.2
No	459	88.8
What is the Source of your information?	(n=58)	%
Friends	4	6.9
Know patient affected with the disease	13	22.4
Reading or university curriculums	31	53.4
Social media	10	17.2
Do you know someone diagnosed with Epidermolysis bullosa?	No. (n=517)	%
Yes	26	5.0
No	491	95.0

As mentioned in Table 1, only 11% of respondents have heard about epidermolysis bullosa disease and the main sources of their information varies between university curriculums (53.4%), knowing patient affected with the disease (22%) or social media (17%). Only 5% who know someone diagnosed with epidermolysis bullosa.

Regarding etiology of epidermolysis bullosa disease and its risk factors, only 8% of respondents were aware about epidermolysis bullosa disease cause, while only 5% of respondents were aware about risk factors of epidermolysis bullosa disease.

As per result, only 3% of participants were aware of epidermolysis bullosa manifestation and the majority respondents (99%) were aware that EB is chronic disease and there is no definitive treatment for it. The overall respondents correct answers regarding to the different complications that may accompany epidermolysis bullosa diseases was only 16%.

Table 2. distribution of respondents according to their perception about seriousness of epidermolysis bullosa disease and currently possible way to prevent it

According to your knowledge of EB disease do you think it is a serious disease?	No. (n=517)	%
Yes	39	7.5
No	478	92.5
What are the possible ways to prevent developing epidermolysis bullosa diseases?	No. (n=517)	%
Avoid marrying a family member	13	2.5
Genetic screening before marriage	38	7.4
Treating affected patients	7	1.4
I Don't know	459	88.8

Table 2 shows 8% of participants agreed upon the seriousness of epidermolysis bullosa diseases and 7% were aware about only current way to prevent epidermolysis bullosa disease.

Surprisingly, when we asked our participants about the importance of adding EB to the premarital screening program, 93.6% of the interviewees stated that adding EB to the screening program is not necessary.

The results of knowledge and awareness assessment implied that only 8% of respondents had good knowledge regarding to epidermolysis bullosa disease, 3% had moderate knowledge, while majority of respondents (89%) had poor knowledge.

DISCUSSION

EP is a rare but detrimental disease. EB is associated with a substantially compromised quality of life that differs according to the disease subtype. The prognosis is excellent for most EB simplex and dominant dystrophic EB patients. Unfortunately, the prognosis is poor for junctional EB and recessive dystrophic EB (Fine et al., 2009; J. D. Fine et al., 2008). Life span of EB patients is also varies according to the subtype of the disease, those with junctional EB are at higher risk of death due to disease complications (J. D. Fine et al., 2008). Since EB is a serious disease and the incidence is rising in Hail city, it is very important to assess the awareness of Hail city population about EP. In our study, we found that most of the participants had poor knowledge regarding EP. A previous study conducted in Rome showed the need to rise the awareness of families of EB diagnosed children (Tabolli et al., 2010). EB puts a huge burden on the family who take care of EB child. Consequently, this huge burden resulted in a broader body surface involvement of blisters and caregiver's greater psychological issues (Tabolli et al., 2010). fortunately, in Hail city, KSA where our study conducted the government take the responsibility of all financial expenses related to the disease which may reduce the burden on the families. Research conducted in Hail city, KSA focused on the genetic causes of dystrophic EB has shown that children with disease belong to the same consanguineous family (Alshammari et al., 2019).Surprisingly, our result showed that most of the participants are against adding EB in premarital screening which support the need of raising the awareness of EB among Hail population. One of the valuable ways to raise awareness among the community is to organize an annual awareness campaign where people can learn more about the disease and understand the impact of EB on the diagnosed children and their families. Poor knowledge is also attributed to the lack of awareness of EB severity among the participants. Therefore, increasing the knowledge of the EB would increase the support of the EB individuals and their families. Nevertheless, increasing the awareness will also promote funding for the essential research for disease treatment and prevention. Nevertheless, no previous studies have been conducted on this topic, neither in KSA nor in other countries, and as far as we know this is the first study to address this issue.

CONCLUSION

EB knowledge was poor among the Hail city population, according to our study. Hail city population and caregivers in particular need to be conscious and educated about EB and its consequences through educational campaign as we mentioned above or through any possible educational ways.

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