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Article

The Significance of the Severity Index of Epidermolysis Bullosa in Newborns in the Conditions of the Fergana Valley of Uzbekistan

Farrukhbek Khamidov[1] , Ayubova Nargiza[2] , M.Z. Khamidova[3]

1. Candidate of Medical Sciences, PhD, Teacher, Department of Dermatovenereology Andijan State Medical Institute, Andijan, Uzbekistan
2. Assistant, Department of Dermatovenerology and Cosmetology Tashkent State Medical University, Tashkent, Uzbekistan
3. Andijan State Medical Institute

*Correspondence: farruh-bek-chamidov@mail.ru[1], n.ayubova@tadhmeduni.uz[2]

Abstract: This study evaluates the significance of the Epidermolysis Bullosa Severity Index in newborns from the Fergana Valley of Uzbekistan. The effectiveness of comprehensive treatment using Colostrum and AVM Neo was assessed. The findings demonstrated a significant reduction in disease severity and confirmed the usefulness of the severity index for diagnosis and treatment monitoring.

Keywords: Epidermolysis bullosa, newborns, severity index, Colostrum, AVM Neo, treatment effectiveness.

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Introduction

Epidermolysis bullosa (EB) is a group of diseases caused by genetic defects and characterized by the spontaneous formation of blisters or their appearance following minor injuries . In recent years, this dermatosis has been diagnosed with increasing frequency in Uzbekistan[1].

According to many researchers, there are three main types of EB:

- simple,
- borderline,
- dystrophic EB,
- as well as their numerous subtypes.

In 2003, Japanese researchers studying rare, intractable skin diseases proposed standards for the diagnosis and treatment of BE. These standards were developed for two types of BE—borderline (LB) and dystrophic (DB). According to the Japanese standards, a diagnosis of juvenile or dystrophic BE is established based on the presence of specific clinical and histological criteria[2].

Clinical criteria:

- Present at birth;

- Blisters form easily on the skin and mucous membranes following minor mechanical irritation;

- Other blistering disorders are ruled out;

- Lesions resolve with atrophy or scarring.

Histological criteria:

Subepidermal (not intraepidermal) blister formation [Table 1][3].

Table 1. Clinical and histological criteria for the diagnosis of BE.

Patient Population	Clinical Criteria	Histological Criteria
Newborn or infant (up to 2 years of age)	1, 2, and 3	Present
Adult	1, 2, 3, and 4	Present

After diagnosing BE based on medical history, cutaneous and extracutaneous symptoms, and the exclusion of other bullous dermatoses, the type of disease is determined. To determine the type, a biopsy of fresh blisters is performed, and the morphological nature of the bulla (intraepidermal, at the dermo-epidermal junction, or intradermal) is established using a light microscope[4]. Japanese authors note that in sporadic cases of EB in newborns and children under 2 years of age, it is virtually impossible to make a clinical differential diagnosis between dominant and recessive dystrophic EB. In such cases, molecular analysis is necessary for an accurate diagnosis[5].

Features associated with juvenile BE: the bulla is located at the dermo-epidermal junction (as shown by electron microscopy); skin atrophy and dental enamel hypoplasia are observed; there are mutations in one of the following genes: laminin 5 α (alpha)-3, β (beta)-3, or γ (gamma)-2 chains; integrin α (alpha)-6 or β (beta)-4 subtype; or the antigen for bullous pemphigoid[6].

Features associated with dominant dystrophic pemphigoid: bullae are located in the dermis (as shown by electron microscopy); both parents and children are affected; a mutation is present in the type VII collagen gene (COL7A1)[7].

Features associated with recessive dystrophic EB [1, 15]: blisters are located in the dermis (as shown by electron microscopy); bullas are observed in a single child or also in siblings (full brothers and sisters); skin scarring and marked syndactyly are present; a mutation in the type VII collagen gene (COL7A1) is present[8].

A Japanese group of BE researchers has proposed several disease severity indices that are important for assessing the prognosis:

1. A separate BE severity index for newborns, since in sporadic cases it is impossible to determine the BE-type dermatosis in newborns;
2. A severity index for borderline EB;
3. A severity index for dystrophic EB.

In addition to the above indices, the Birmingham Epidermolysis Bullosa Severity Score (BEBS) is also used to diagnose EB[9].

The aim of the study is to investigate the “Severity Index of Epidermolysis Bullosa in Newborns” in newborn patients with EB undergoing comprehensive therapy with the

dietary supplement Colostrum (colostrum) and AVM Neo in the Fergana Valley, Uzbekistan.

Materials and Methods

We observed 25 newborns aged 1 to 15 days (13 boys, 12 girls). The "neonatal epidermolysis bullosa severity index" was assessed in all patients. Patients with EB received standard treatment in combination with the dietary supplement "Colostrum" (colostrum) and AVM Neo. Colostrum was administered orally at a dose of 0.5 mL twice daily before meals for 1 month. Affected children took AVM Neo at a dose of 1 teaspoon 2–3 times a day for 30 days[10].

The neonatal epidermolysis bullosa severity index includes 6 main and 5 supplementary parameters [Table 2].

The primary parameters include:

- the appearance of new bullae,
- the extent of bullae (%),
- bullae in the oral cavity,
- feeding difficulties,
- weight changes,
- staphylococcal infection,
- methicillin-resistant.

Additional parameters include:

- laminin 5 deficiency,
- pyloric atresia,
- family history of a fatal case of BE,
- presence of severe recessive dystrophic BE in the family history,
- a family history of BE with muscular dystrophy.

Table 2. Calculation of the BE severity index in newborns[11].

Parameters	Indicators	Score
New bullae	None	0
	1–4 per week	1
	5–10 per week	2
	>10 per week	3
Area of blister distribution	None	0
	1–4%	1
	5–15%	2
	>15%	3
Bullae in the oral cavity	None	0
	1–2 per month	1
	Several per month	2
	Persistent, always present	3
Feeding difficulties	None	0
	Mild	1
	Moderate	2

	Severe	3
Weight changes	Normal weight gain	0
	Low weight gain	1
	No weight gain	2
	Weight loss	3
Methicillin-resistant *Staphylococcus aureus* (MRSA) infection	None	0
	Localized, inactive infection	1
	Localized, active infection	2
	Systemic infection	3
The total score is calculated based on all six parameters.		

Interpretation of the index (The maximum score is 18; a score of 0 to 5 indicates a mild condition, 6 to 10 indicates a moderate condition, and 11 to 16 indicates a severe condition; the patient is classified as severe if they have 1 or more points plus the presence of at least one of five additional criteria (laminin 5 defect, pyloric atresia, family history of a fatal case of BE, a family history of severe recessive dystrophic BE, or a family history of BE with muscular dystrophy))[12].

Results and Discussion

Prior to treatment, the "Neonatal Epidermolysis Bullosa Severity Index" in newborns with EB averaged 6.35 ± 2.12 points. After comprehensive therapy with Colostrum and AVM Neo, the "Neonatal Bullous Epidermolysis Severity Index" in newborns with BE decreased to 2.03 ± 0.19 points[13]. As the study results show, in patients with epidermolysis bullosa, the use of Colostrum and AVM Neo significantly reduced the "Newborn Bullous Epidermolysis Severity Index." The use of these medications as part of a comprehensive treatment regimen has a sufficiently effective impact on skin healing and shortens the duration of treatment in the Fergana Valley region of Uzbekistan[14-15].

Conclusion

The "Newborn Bullous Epidermolysis Severity Index" aids in the accurate diagnosis and assessment of the effectiveness of BE treatment in newborns in the Fergana Valley, Uzbekistan. Furthermore, the medications Colostrum and AVM Neo improve the effectiveness of standard treatment for this dermatosis and provide grounds for their inclusion in the comprehensive treatment of EB.

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