The efficacy of nutritional screening tools for diagnosis of malnutrition in children with epidermolysis bullosa

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Abstract: Malnutrition worsens the course of the main disease and requires early diagnosis. We were aiming to identify the prevalence of malnutrition and to elicit the most effective nutritional screening tool for malnutrition in children with epidermolysis bullosa. A cross-sectional study for prevalence and suitable nutritional screening tool for malnutrition of 26 patients of age 2 to 18 years with mean age 8.65±3.86 were carried out. There were 14 females (53.8%) and 12 males (46.2%). Anthropometric data, medical and nutrition history were obtained. The following nutritional screening tools were studied: PYMS, STAMP, STRONGkids, and we calculated the degree of malnutrition with online calculator PediTools, taking into account Z-score of anthropometric indicators. We evaluated the validity of these screening tools by calculating the sensitivity and specificity alongside their accuracy with the 2x2 table using MEDCALC statistical software. According to clinical manifestation, there were 11 (42.3%) children with severe malnutrition, 7 (26.9%) with moderate and 8 (30.8%) without malnutrition. Using different nutritional screening tools we found the following: on STRONGkids, 14 (53.9%) had low risk, 3 (11.5%) - moderate risk, 9 (34.6%) - high risk of malnutrition, sensitivity was 66.67%, specificity - 100%, accuracy – 76.92%. On STAMP we found, with 5 (19.2%) low risk, 7 (26.9%) - moderate risk, 14 (53.9%) - severe risk, sensitivity was 100%, specificity – 62.5%, accuracy – 88.46%. On PYMS, there were 11 (42.3%) with low risk and 8 (30.8%) with moderate risk, 7 (26.9%) with severe risk; sensitivity was 83.33%, specificity - 100%, accuracy – 88.46%. We did not find a good nutritional tool for screening of malnutrition in patients with epidermolysis bullosa. But PYMS have shown more efficiency in comparison with STRONGkids and STAMP because it includes evaluation of BMI which makes it possible to evaluate whether the body mass is insufficient or normal.

Keywords: epidermolysis bullosa, nutrition assessment, nutritional status, malnutrition, pediatrics.

Introduction
In hospitalized patients, malnutrition is a significant yet hidden problem that can determine the disease outcome, hence the need for nutritional screening tools and proper nutritional care as nutritional involvement is directly related to the severity of the associated clinical complication. Marta Gambra-Arzoz et al estimated in a study with 282 hospitalized children with different pathology that the prevalence of acute and chronic malnutrition is 13.7% and 7.4% respectively (Gambra-Arzoz et al., 2020).

Other recent studies estimated 12-24% as malnourished in hospitalized children (Shaughnessy et al., 2021).
Epidermolysis bullosa (EB) is a group of inherited rare genetic dermatoses characterized by mucocutaneous fragility and blister formation, often triggered by minimal skin friction. The main types of EB are EB simplex, junctional, dystrophic and Kindler (Bardhan et al., 2020).

EB can be inherited as autosomal dominant or recessive with varying degrees of severity ranging from mild to fatal. The major symptoms of any type of EB include scars, hair loss and deformities of extremities, fragile skin with blisters generalized or localized on the skin and mucosa. It also involves extracutaneous complications such as esophageal strictures, squamous cell skin cancer, constipation, malnutrition, gastroesophageal reflux disease, anemia etc. (Mariath, Santin, Schuler-Faccini & Kiszewski, 2020).

Nutritional deficiency can develop from effects of extracutaneous involvement and increased burden on the body metabolism and immune system complication the disease itself creating a vicious cycle of complications. Also, extracutaneous blistering and narrowing of the esophagus will decrease food intake and further decrease the available nutrient for the highly stressed body. All these factors contribute to the development of malnutrition in EB patients (Zidorio et al., 2015).

Nutritional screening tools are important to identify the risk of malnutrition on admission but there are several nutritional screening tools (NST) and the question remains, how does one choose from this pool of available nutritional screening tools being that each tool has been designed with different goals and application process?

Some papers in the literature identified STRONGkids as an easy-to-use nutritional screening tool for hospitalized patients (Hulst, Zwart, Hop, & Joosten, 2010; Joosten & Hulst, 2014; Durakbaş, Fettahoğlu, Bayar, Mutus & Okur, 2014), however, STRONGkids showed low efficiency in identifying the risk of malnutrition in some diseases (da Cruz Gouveia, Tassitano & da Silva, 2018). Hence, the need for eliciting the most effective tool for identifying the risk of malnutrition in diseases like EB.

According to the European Society of Parenteral and Enteral Nutrition (ESPEN), the use of NST can help with early recognition preventing further deterioration in hospitalized patients thereby reducing their hospital cost and stay time (Hartman, Shamir, Hecht & Koletzko, 2012; Woonputri, Djais & Rosalina, 2014).

The available NST for pediatric patients include:
1. Nutritional Risk Score (NRS)
2. Pediatric Nutritional Risk Score (PNRS)
3. Screening Tool for the Assessment of Malnutrition in Pediatrics (STAMP)
4. Subjective Global Nutritional Assessment (SGNA)
5. Pediatric Yorkhill Malnutrition Score (PYMS)
6. Screening Tool for Risk Of impaired Nutritional Status and Growth (STRONGkids)

**Aim**

The aim of our study was to identify the prevalence of malnutrition in EB patients among different age groups and to elicit the most effective Nutritional Screening Tool for the predicting of malnutrition in children with EB.

**Materials and Methods**

**Patients**

A cross-sectional study for prevalence and suitable nutritional screening tool for malnutrition of 26 EB patients of age 2 to 18 years was carried out at the EB cabinet in Ukrainian National Children’s Specialized Hospital «OKHMAT-DYT».

For this research, we compared the efficiency of STRONGkids, STAMP, PYMS in predicting the risk of malnutrition.

**Data collection**

Patients’ general data parameters were collected and they include the date of birth, age, sex, anthropometric parameters included height, weight, body mass index (BMI), recent and unintentional weight loss, gastrointestinal complaints.

**Reference Standard**

We compared the results of the selected Nutritional screening tools to the anthropometric Z-scores of these EB patients. We obtained each
Patient's Z-score using the online calculator Pediatric Tools which allowed us to calculate weight, height and BMI, as well as exact percentile and Z-score based on the Center for Disease Control growth charts.

It is widely accepted that the normal nutritional value of Z-score is above -2.0 SD; moderately malnourished if Z-score of two anthropometric parameters (BMI, or weight for age, or weight for height (WFH), or height for age) are between -3.0 and -2 SD; severely malnourished if Z-score is below -3.0 SD or edema (Bouma, 2017).

**Screening Tool for Risk of Impaired Nutritional Status and Growth (STONGkids)**

STONGkids is a nutritional screening tool designed to identify the risk of malnutrition and was developed according to the ESPEN guidelines. (Huysentruyt et al., 2013). It comprises of four questions on subjective clinical assessment, high-risk disease, feeding characteristics, weight loss or poor weight gain. Each question is scored 1-2 points with a maximum total score of 5 points. Adding all scores assesses the total risk of malnutrition. High risk malnutrition is recorded when total risk = 4-5 points, moderate risk =1-3 points, low risk=0 point.

**Screening Tool for the Assessment of Malnutrition in Pediatrics (STAMP)**

A team from Royal Manchester Children’s Hospital and the University of Ulster, the lead investigator being Helen McCarthy, Lecturer and Honorary Pediatric Dietitian, developed STAMP (www.stampscreeningtool.org). It consists of 3 elements: Diagnosis with nutritional implications, Nutritional intake and Child’s measurement (height and weight). For this purpose, we used the centile quick reference table according to the Center for Disease Control. The total score was labeled as high risk when total risk ≥4 points, medium risk =2-3 points, low risk = 0-1 points.

**Pediatrics Yorkhill Malnutrition Score (PYMS)**

PYMS was created in Glasgow according to a national standard set that identified the importance of screening for malnutrition in patients over 1 year old. It consists of 4 questions: about recent weight loss, reducing food intake, possible affection of nutrition due to recent hospitalization and estimation of BMI in children. The high risk of malnutrition is determined when the total risk is ≥ 2 points.

**Statistical analysis**

We analyzed all data using Microsoft Excel. The results are presented as the mean and standard deviation (M±SD). The test for normality of distribution of the sample was carried out by the Kolmogorov-Smirnov test.

We evaluated the validity of PYMS, STAMP, STONGkids, by calculating the sensitivity and specificity alongside their accuracy with the 2x2 table using MEDCALC statistical software. Confidence interval (CI 95%) was determined separately for each of these parameters. P-value less than 0.05 was considered statistically significant.

**Results**

We analyzed a total of 26 patients suffering from EB. The patients aged between 2 to 18 years with mean age 8.65±3.86. Most patients (65.4%) had the dystrophic type of EB (Table 1). 69.2% of EB patients had chronic protein-energy malnutrition of moderate and severe degree and only 30.8% were with normal nutritional status (fig. 1). Most of the children with DEB suffered

![Fig. 1. Distribution of the patients with EB due to severity of malnutrition](image-url)

<table>
<thead>
<tr>
<th>Table 1. The Demographics characteristics of the patients with EB</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Patient characteristics</strong></td>
</tr>
<tr>
<td>Age (years)</td>
</tr>
<tr>
<td><strong>Sex frequency</strong></td>
</tr>
<tr>
<td>Female</td>
</tr>
<tr>
<td>Male</td>
</tr>
<tr>
<td><strong>Types of EB</strong></td>
</tr>
<tr>
<td>Simplex</td>
</tr>
<tr>
<td>Junctional</td>
</tr>
<tr>
<td>Dystrophic</td>
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</tbody>
</table>
problems with swallowing (dysphagia) and only 18.2% of patients with severe malnutrition had a similar symptom. In our view, children with severe malnutrition do not complain very often on dysphagia due to the predominant use of liquid food in this group (most of the patients in this group had dilatation of esophagus due to stenosis) in contrast to 57.1% of patients with moderate malnutrition who eat solid and liquid food and subjectively complaining about problems with swallowing. All described data are presented in Table 2.

Evaluation of STAMP, PYMS AND STRONGkids tools in EB patients

Using different nutritional screening tools, we found the following:

On STRONGkids, 14 (53.9%) had low risk, 3 (11.5%) - moderate risk, 9 (34.6%) - high risk, sensitivity was 66.67%, specificity - 100%, accuracy – 76.92%.

On STAMP we found, with 5 (19.2%) low risk, 7 (26.9%) - moderate risk, 14 (53.9%) - severe risk, sensitivity was 100%, specificity - 62.50%, accuracy - 88.46%.

On PYMS, there were 11 (42.3%) with low risk and 8 (30.8%) moderate risk, 7 (26.9%) with severe risk, sensitivity was 83.33%, specificity - 100 %, accuracy - 88.46%.

These variables are shown in Table 3.

With the 2x2 table using MEDCALC statistical software we calculated the validity that is shown in Table 4.

We did not find a good nutritional tool for screening malnutrition in EB patients. Sensitivity for STAMP and PYMS were 100% and 83.33% respectively.

We found more efficiency with PYMS being the most efficient in the prediction of malnutrition in EB patients in comparison with STRONGkids, which showed 66.67% sensitivity.

Table 2. Prevalence of gastrointestinal complaints in EB patients with different severity of malnutrition

<table>
<thead>
<tr>
<th>COMPLAINTS</th>
<th>DEGREE SEVERITY OF MALNUTRITION</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Normal</td>
</tr>
<tr>
<td>Dental problems</td>
<td>62.5%</td>
</tr>
<tr>
<td>Chewing problems</td>
<td>37.5%</td>
</tr>
<tr>
<td>Constipation</td>
<td>37.5%</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>25.0%</td>
</tr>
<tr>
<td>Get full quickly</td>
<td>12.5%</td>
</tr>
<tr>
<td>Nausea</td>
<td>25.0%</td>
</tr>
<tr>
<td>Vomiting</td>
<td>12.5%</td>
</tr>
</tbody>
</table>

Table 3. The results of nutrition screening tools in EB patients

<table>
<thead>
<tr>
<th>NST</th>
<th>STAMP</th>
<th>STRONGkids</th>
<th>PYMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low</td>
<td>5 (19.2%)</td>
<td>14 (53.9%)</td>
<td>11 (42.3%)</td>
</tr>
<tr>
<td>Moderate</td>
<td>7 (26.9%)</td>
<td>3 (11.5%)</td>
<td>8 (30.8%)</td>
</tr>
<tr>
<td>Severe</td>
<td>14 (53.9%)</td>
<td>9 (34.6%)</td>
<td>7 (26.9%)</td>
</tr>
</tbody>
</table>
Discussion

Most works in literature recommended STRONGkids as a more efficient nutritional screening tool for patients with chronic diseases (Huysentruyt et al., 2013; Durakbaş et al., 2014; Lara-Pompa et al., 2020), but with a focus on EB patients, STRONGkids was very inefficient. This is because EB is not included in the disease list for calculating STRONGkids. Also, STRONGkids focuses on disease symptoms occurring in the last 1-3 days and a few weeks to months, which does not favor patients with chronic diseases occurring for years like in EB.

Undernutrition is classically subdivided into acute undernutrition (defined by the World Health Organization (WHO) as the weight for height [WFH] <-2 SD) and chronic undernutrition (defined by the WHO as height for age [HFA] <-2 SD).

Koen Huysentruyt et al were the first to validate the use of the STRONGkids by nurses as an easy-to-use rapid nutritional screening tool for hospitalized patients. In their study with the Belgian population of hospitalized children they found a good correlation of STRONGkids with acute malnutrition, but not with chronic undernutrition. (Huysentruyt et al., 2013). This means that STRONGkids has a poor correlation with chronic undernutrition, which further reinforces our result.

Lama More RA et al studied nutritional screening tools for hospitalized pediatric patients in Spain and they tried to validate STAMP for nutritional screening (Lama More et al., 2012). They concluded that STAMP is a simple and useful tool for nutritional screening, avoiding the need to assess all patients on admission in order to identify those under nutritional risk.

Rebecca Ling et al compared STAMP with STRONGkids and concluded that STRONGkids was superior to STAMP in terms of validity because STAMP over-diagnosed the number of children with nutritional risk and STRONGkids had a closer correlation to the nutritional status but this does not apply to patients with EB and it further reiterates the fact that each screening tool has been designed with different goals (Rebecca, Victoria Hedges & Peter, 2011).

In our study, STRONGkids under-diagnosed the number of children with nutritional risk. STRONGkids found 53.9% of the patients as low risk, 11.5% as moderate risk, and 34.6% as high risk and this does not correlate with the nutritional status recorded by Z-score as 42.3% with severe malnutrition, 26.9% moderate, and 30.8% with normal malnutrition.

PYMS-assessed nutritional risk showed the closest correlation with Malnutrition status in accordance to Z-score and this was also confirmed by the high results of 83.33% Sensitivity, 100% specificity, 88.46% accuracy followed by STAMP which showed 100% sensitivity, 62.50% specificity, 88.46% accuracy.

STRONGkids did not show a strong correlation to the standard nutritional status.

There is no universally accepted nutritional screening tool for predicting the risk of malnutrition but this is mainly due to the lack of a universally accepted definition for malnutrition. EB is a rare genetic disease but the need to identify malnutrition and the risk of malnutrition cannot be overemphasized as this determines the effectiveness of its management.

Malnutrition is associated with many outcomes including longer hospital stay, increase morbidity and mortality and increase cost of disease management.

There are six nutritional screening tools for hospitalized pediatric patients and all six can

### Table 4. Evaluation of STAMP, PYMS AND STRONGkids tools in EB patients

<table>
<thead>
<tr>
<th>Index</th>
<th>STAMP</th>
<th>STRONGkids</th>
<th>PYMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity, 95% CI, %</td>
<td>100.00 %</td>
<td>66.67 %</td>
<td>83.33 %</td>
</tr>
<tr>
<td>Specificity, 95% CI, %</td>
<td>62.50 %</td>
<td>100.00 %</td>
<td>100.00 %</td>
</tr>
<tr>
<td>Accuracy, 95% CI, %</td>
<td>88.46 %</td>
<td>76.92 %</td>
<td>88.46 %</td>
</tr>
</tbody>
</table>

Table 4. Evaluation of STAMP, PYMS AND STRONGkids tools in EB patients
identify the risk of malnutrition but not accurate in all diseases and for this reason we compared the efficiency amongst STAMP, PYMS, and STRONGkids.

Against the popular notion in the literature that STRONGkids can be the most effective tool for identifying the risk of malnutrition, we found PYMS to be the most sensitive followed by STAMP.

Epidermolysis Bullosa is a chronic lifetime disease, therefore, a screening tool that focuses on eliciting acute symptoms will not be effective as seen with STRONGkids.

Nutritional screening should be conducted on admission and on regular follow-up assessment during admission (Joosten & Hulst, 2014).

Conclusions
In conclusion, 69.2% of EB patients are suffering from chronic protein-energy malnutrition moderate or severe degree. We do not have a gold standard nutritional tool for effective identification of malnutrition in EB patients, but PYMS and STAMP have shown a positive correlation with reference nutritional status while STRONGkids is poorly efficient for EB patients.

Recommendation
We recommend that PYMS and STAMP nutritional screening tools be studied in a larger EB population as they show a more prospective result for calculating the risk of malnutrition in EB patients. Taking into consideration the particulars of the disease in our patients, it would probably be better to develop a specific nutritional screening tool for EB patients, which would take into account the most common gastrointestinal complications that affect the development of malnutrition.

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A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of article

REFERENCES


Ефективність скринінгових опитувальників для діагностики мальнутриції у дітей із бульозним епідермолізом

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Анотація. Білково-енергетична недостатність (мальнутриція) погіршує перебіг основного захворювання, тому потребує ранньої діагностики. Метою нашого дослідження було встановити частоту білково-енергетичної недостатності (БЕН) у дітей із бульозним епідермолізом (БЕ) та дослідити ефективність скринінгових опитувальників для визначення її ризиків виникнення. Було проведено одномоментне дослідження, в якому брали участь 26 пацієнтів з БЕ віком від 2 до 18 років (середній вік 8,65±3,86 р.). Серед пацієнтів було 14 (53,8%) дівчат та 12 (46,2%) хлопців. Пацієнтам було проведено антропометрію та оцінено нутритивний статус. Для визначення ризику розвитку БЕН були використані такі скринінгові опитувальники: PYMS, STAMP, STRONGkids. Ступінь тяжкості БЕН визначали, враховуючи Z-критерій антропометричних показників, за допомогою онлайн калькулятора PediTools. Ми оцінили валідність скринінгових опитувальників, визначивши чутливість, специфічність та точність за допомогою таблиці 2x2 статистичного програмного забезпечення MEDCALC. Серед обстежених пацієнтів із БЕ тяжку БЕН було діагностовано у 11 (42,3 %), БЕН помірної важкості – у 7 (26,9 %), а в 8 (30,8 %) дітей ознак БЕН не було виявлено. За результатами опитувальника STRONGkids 14 (53,9%) дітей мали низький ризик БЕН, 3 (11,5%) дітей мали помірний ризик та 9 (34,6%) дітей – високий ризик, чутливість склали 66,6%, специфічність – 100 %, точність – 76,92%. За результатами опитувальника STAMP ми виявили у 5 (19,2%) дітей низький ризик БЕН, у 7 (26,9%) – помірний ризик, у 14 (53,9%) – високий ризик, чутливість склали 100%, специфічність – 62,5%, точність – 88,46%. За результатами опитувальника PYMS було 11 (42,3%) дітей з низьким ризиком БЕН, 8 (30,8%) з помірним ризиком, 7 (26,9%) з високим ризиком; чутливість – 83,33 %, специфічність – 100 %, точність – 88,46 %. Ми не знайшли ідеального скринінгового опитувальника для визначення ризику розвитку БЕН у пацієнтів з бульозним епідермолізом. Але PYMS показав більшу ефективність у порівнянні з STRONGkids та STAMP, оскільки він включає оцінку IMT, що дає змогу опосередковано судити про нутритивний стан дитини.

Ключові слова: бульозний епідермоліз, оцінка харчування, стан харчування, мальнутриція, педіатрія.