UDK 615.851.3:616.831-009 **Professional Paper**
doi: 10.7251/SANUS2401249Z

OCCUPATIONAL THERAPY PROCESS IN EPIDERMOLISIS BULLOSA

Edina Pulić[1], Manuela Komorski[2], Jelena Klanjčić[3], Tatjana Njegovan Zvonarević[1], Maja Trulec[4]
[1]University of Applied Health Sciences, Mlinarska cesta 38, Zagreb, Croatia;
[2]Croatian Association of the Deaf-Blind Dodir, Medulićeva 34, Zagreb, Republic of Croatia
[3]Special hospital for chronic diseases of childhood Gornja Bistra, bolnička ulica 21, Gornja Bistra, Republic of Croatia
[4]Debra, Society of Epidermolysis Bullosa Patients, Prilaz Gjure Deželića 30, Zagreb, Republic of Croatia

Abstract. *Epidermolysis bullosa (EB) belongs to a group of rare, genetic diseases characterized by extreme sensitivity of the skin and in some forms of the mucous membrane. The progressive nature of this disease disrupts the life of the affected person and gradually leads to disability. It is a hereditary disease in which the slightest pressure or friction can cause painful blisters and wounds. Complications can also occur on the mucous membranes of the eyes, mouth, and digestive system. Thise paper presents an overview of the impact of EB on quality of life, the connection between occupations and health, the occupational therapy (OT) process for persons with EB, and the role of support systems and intersectoral cooperation. Emphasis is placed on the progression, challenges, and impact of the disease on the daily life of persons with EB from birth to adulthood. Accordingly, OT interventions are also presented, the primary goal of which is to overcome obstacles and strengthen occupational participation as a means for improving the quality of life of persons with EB. Thise paper provides insight into the complexity of this area, taking into account research in the field of OT practice and the quality of life of persons with EB.*

Key words: *epidermolysis bullosa, occupational therapy process, quality of life, occupational participation*

Introduction

Epidermolysis bullosa (EB) is a very rare and severe genetic disease that affects the largest human organ - the skin, but also the mucous membrane. It is characterized by the fragility of the skin with the appearance of blisters spontaneously or after minor trauma such as gentle pressure or friction. There are four main types of EB: EB Simplex (EBS), Junctional (JEB), Dystrophic (DEB) and Kindler syndrome (KS). Broadly speaking, EBS encompasses all subtypes of EB caused by mechanical fragility and blistering limited to the epidermis. JEB includes all subtypes with bleb formation within the basal lamina and basement membrane. In DEB, blisters are formed in the uppermost layer of the dermis, and in KS, the formation of blisters occurs at multiple levels within or below the basement membrane. Table 1 shows the clinical findings of the latest international consensus on the diagnosis and classification of EB in 2014 [1]. The table summarizes the relative occurrence of aspects of EB that may affect function within the various subtypes.

Table 1. Relative occurrence of aspects of EB that may affect function within various subtypes [1].

	EB simplex	Junctional (JEB)	Dystrophic (DEB)	RDEB
Blisters	4+	2-4+	2-3+	3-4+
Atrophic scars	Rare -2+	Not present - 3+	3-4+	3-4+
Relative incidence of blisters	Frequent	2+-4+	Variable	High
Pseudosyndactyly	Not present	Not present - 1+	Rare	Variable -4+
Explanation: relative frequency; absent or none; rare; 1+;2+;3+;4+, EB: Epidermolysis bullosa; DDEB: dominant dystrophic EB; ERDEB: recessive dystrophic EB.				

EB belongs to the group of non-infectious skin diseases. According to unofficial data from the Croatian Association for Rare Diseases, there are 60 families with a sick member in Croatia, but there is still no register that would ensure the exact number of sick people. On a national basis, there is no precise guideline on the reporting of non-infectious skin changes, which is why little is still known about the epidemiology of this disease. It occurs in two to four people per 100,000 inhabitants of all races and ethnic groups, and affects both sexes equally [2]. The disease is most often manifested at birth or in early childhood, and very rarely later in life [3], it is inherited in an autosomal dominant or autosomal recessive manner. People with EB have a range of symptoms such as scarring, fibrosis and contractures affecting any part of the body. Pain (acute, chronic) that affects independence in activities of daily living (ADLs) and also the quality of life (QoL) often occurs. Chronic blistering on the skin and mucous membranes, pain, itching, unpleasant odor and long-term bandages are just some of the problems that affect the personal, physical, emotional and professional aspects of the life of the affected person. There is currently no cure for EB, and treatment is carried out with proper skin care, regular medical examinations, dietary adjustments and surgical procedures. The aim of this paper is to describe the development and implementation of an occupational therapy plan and interventions that are adapted to the individual needs of people with EB with the aim of improving quality of life, increasing functional independence through inclusion in occupations, reducing pain and the risk of injuries, and improving psychosocial well-being. Also, this work aims to clearly define OT evaluation and intervention for people with EB which includes habilitation and rehabilitation approaches as well as support and empowerment of parents so that they can effectively care for their child and improve the quality of life of the whole family.

Impact of EB on quality of life

QoL is a multidimensional concept, it is difficult to define, that is, there is no one generally accepted definition. QoL is an abstract, multidimensional construct that reflects the physical, psychological and social aspects of an individual's well-being. It represents an important clinical outcome, therefore it is the focus of many scientific disciplines. The World Health Organization (WHO) defines it as "Individual perception of one's own life position in the context of culture and value system in relation to one's own goals" [4]. Assessment of QoL is complex and includes non-clinical variables such as personality, ambitions, expectations, socioeconomic level, marital status, religious experience, and other factors [5]. For the purposes of assessing the QoL of people with EB, Frew et al. developed the Quality of Life in Epidermolysis Bullosa (EB-QoL) questionnaire, which encompasses the physical and emotional impact of EB on the personal and social life of patients, as well as their productivity at work and school [6]. Accordingly, this questionnaire has proven to be a useful tool for assessing the QL of patients with EB and should be used in RT studies of QoL. EB has a significant clinical and socioeconomic impact on patients and their families [7], therefore a general approach to assessing the QoL of families or caregivers is needed to assess the burden of the disease. People with EB can have different clinical symptoms. The most common symptoms are blisters, ulcers, skin infections, pain, pruritus, esophageal stricture, microstomia, ankyloglossia, dysphagia, gastrointestinal ulcers, malnutrition, pseudosyndactyly, and scarring leading to contractures. The mentioned symptoms represent physical symptoms and are the main cause of limitations in the performance of ADLs [8-10] and significantly affect QoL [11]. Wounds and blisters, pseudosyndactyly, and poor joint mobility affect mobility, grips, feeding, and self-care in general. Carrying out daily occupations can cause painful wounds that are difficult to heal and cause constant formation of new ones. Wounds and blisters can also affect the mucous membrane of the eye and lead to tearing and pain, and sometimes to permanent damage to vision, which in turn leads to difficulty in movement and performance of daily occupations [2]. Involvement of the mucous membrane of the mouth and esophagus (joining of the tongue, narrowing of the esophagus, difficult opening of the mouth, poor quality and/or lack of teeth) leads to difficulty maintaining oral hygiene, feeding and digestion problems. Feeding activity is very demanding for people with EB because it leads to fatigue, difficult feeding, malnutrition, anemia, constipation and poor wound healing. The aforementioned physical factors affect numerous life segments such as play, education, employment, sociability and participation in the community [9]. Physical factors can result in depression and anxiety [13]. From the moment of birth, patients feel their first experiences (touches, tastes and steps) through pain, and because of this, everyday challenges are manifested through negative emotions (fear, sadness, anger, dissatisfaction, frustration). These negative emotions reflect on self-confidence and self-esteem and lead to a loss of interest and motivation. Many patients become inactive in ADLs, they go out into the community less, which results in impaired socialization and social isolation. Non-participation in desired activities and occupations leads to occupational imbalance and deprivation, which additionally affects health and well-being. Everyday life with numerous obstacles such as parents' fear and anxiety often result in the child not being included in educational institutions, which later affects his level of independence and the possibility of employment. Unemployment, reduced income, dependence on the help of others and non-involvement in activities prevent sick people from being full members of the community.

Connection between occupation and health

The fundamental belief of the OT profession is the positive relationship between occupations and health and the perception of people as occupational beings [14]. Occupations are activities that people do every day to give meaning and purpose to their lives. They can be done independently, with family members and friends, at home, at work or school, and in the community. Without involvement in occupations, people do not have the opportunity to act, create, enjoy, learn, develop and socialize [15]. Mayer, one of the founders of the OT profession as well as the Ottawa Charter for Health Promotion emphasize the strong connection between occupations, health and the environment [16]. The connection between occupations and health is particularly visible in situations of illness, injury or developmental difficulties when participation in occupations is impaired. The impossibility of fulfilling occupational needs has a negative effect on the daily life of the patients, on their family dynamics and sociability [17] and on the QoL. The imperative of the RT profession is focused on health, not only on occupational performance and participation, and includes interventions within the physical, social, mental, emotional and cultural aspects of action, existence, longing and belonging. RT Vision 2025 states that "Occupational therapy improves the health, well-being and quality of life of all people, populations and communities through participation in everyday life" [16].

Occupational therapy process in EB

Evidence-based practice has indicated the need for clinical practice guidelines to establish the foundation of occupations, create and improve standards of care, and effective OT interventions in working with EB patients. In 2019, OT guidelines for working with EB patients were defined [18]. *The OT process begins with an assessment*. It is recommended that patients with all subtypes of EB with functional or biomechanical impairments, including contractures and reduced mobility, undergo an assessment of functional independence in ADLs as soon as possible with frequent repeated assessments. The OT Evaluation of EB - Occupational Therapy Evaluatin for Epidermolysis Bullosa is a specially designed evaluation that is used exclusively in working with people with EB. By carrying out the assessment, general information about the sick person, information related to the diagnosis (form of the disease, drugs used) and the social environment is obtained. Self-care activities, productivity activities, person's mobility and the rhythm of wakefulness and sleep are evaluated and scored. A special feature of the assessment is the segment related to determining and analyzing the function of the hands. Hand dominance is identified, functional grips are assessed, and finger distances are measured to monitor the fusion process and current range of motion within the hand [13]. In addition to the above assessment, the Canadian Occupational Performance Measure (COPM, Law et al., 1991), the

Barthel Index (BI) (Mahoney and Barthel 1965), can also be used in working with people with EB. Safety Assessment of Function and the Environment for Rehabilitation - SAFER, Chiu, Oliver, Marshall and Letts, 1993) and Child Occupational Self-Assessment - COSA, Keller, Kafkes, Basu, Federico and Kielhofner, 2005). The COPM is a standardized, clinically proven, evidence-based assessment that assesses the outcomes of an intervention over a period of time from the unique perspective of the individual. Its effectiveness has been demonstrated in interdisciplinary community rehabilitation programs [19-21], in improving patient-centered care [22], facilitating patient-centered goals, and assessing outcomes [23-25]. The Barthel Index is an ordinal scale that measures a person's ability to perform ADLs. It was originally created to assess disability in patients whose impairments affect the use of limbs when performing ADLs [26]. SAFER is used in assessing the safety of the home, living conditions, assessing the environment as well as the performance capabilities of the person who lives in it. COSA assesses children's sense of competence in participating and completing activities and the value of these activities [27]. *OT intervention* in EB includes habilitation and rehabilitation. Immediately after the birth of a child with EB, it is necessary to include the child and parents in early rehabilitation. It is recommended that patients are involved in defining therapeutic goals appropriate to their age throughout their entire lifespan. One of the leading and most frequent problems that appear are contractures of the hands and feet, which lead to further damage and difficulties in performing basic ADLs such as dressing, feeding and bathing. The OT intervention is aimed at promoting independence in ADLs, which should be integrated into everyday family life. It may include choosing appropriate clothing, adjusting the environment, for example, toothbrushes, equipment needed for bathing, toileting, etc. *OT intervention in babies and young children*. The focus of the RT process in this phase is aimed at identifying the needs of parents and providing support that will help them ensure the quality upbringing and development of their child. It is important to educate parents about "baby handling" and about the methods and adaptations they can use. A child with EB should be approached carefully and held in an adapted way to prevent skin friction and the formation of new wounds. First of all, it is necessary to determine where the child has wounds on their body so that they can be approached in the most adequate and painless way. It is recommended to lift the child so that one hand is placed under the buttocks and the other in the neck area rather than lifting the child in the armpit area. Soft materials that do not irritate the skin can also be used to lift the child. It is recommended that clothes and shoes are made of silky textures and a simple design that is easy to wear. It is not recommended to bathe a child every day because it is very difficult and tiring. The diapers used should be disposable and made of soft materials with edge protection to prevent skin friction and the formation of new wounds [28]. Socks and gloves are used to prevent the infant from rubbing their hands and feet and from scratching their face to prevent new skin damage. During breastfeeding, it is possible to damage the skin on the child's face, so it is recommended to apply Vaseline on the mother's chest. The activity of feeding can be made more difficult by the appearance of mouth sores, so the choice of cutlery material is extremely important. Bottles and spoons with a soft texture are used, usually silicone [29]. Children should be encouraged to explore their surroundings, which is a lifelong skill that should be started as early as possible. When the child begins to crawl, it is recommended to put soft protections with a slight fixation on the bony segments of the hands, elbows, hips and knees. All of the above will protect the skin from the appearance of blisters and the creation of additional damage. *OT intervention in the preschool period*. Enrollment in an educational institution is the first step towards a child's greater independence and the development of various skills, especially social skills that are key to building character, self-confidence, self-regulation, empathy and improving communicationWhen enrolling a child in kindergarten, education and preparation of the environment in which the child comes is important. The OT process in kindergarten age is based on play, which is recognized as an effective OT medium in working with children and their families. In early childhood, play is the driving force of learning, through it the child acquires knowledge, develops skills and speech, improves communication, and thus socialization. They use the OT game as a means to achieve numerous development goals. Children with EB should be allowed to participate in all forms of play and adapted to the child's capabilities. When choosing a toy, it is necessary to take into account the size, composition and texture of toys without sharp edges and rough materials that could injure the child's sensitive skin [9]. Besides playing, writing, drawing and painting are one of the most important occupations of children. To perform the mentioned occupations, the functionality of the hands and numerous grips are needed, which in all people begin to develop and form very early, but in patients with EB this development is limited. The development of grasping skills, manipulation of objects and graphomotor skills comprise a large part of the OT intervention in this period of life [13]. OT intervention for grips includes individual wrapping of the fingers and the use of thermoplastic orthoses with or without silicone inserts, continuous activation of muscles and joints that will delay contractures and deformations and improve functional mobility. When painting and using art materials, it is important to protect the child's hands so that the wounds on the hands do not become infected. Here, special cotton gloves intended for children with EB help prevent itchy skin, protect the skin from infection, and maintain a certain position of the fingers to slow down the fusion process [9]. It is also necessary to advise parents about all the benefits and contraindications of wearing gloves, emphasizing that it is not desirable for the child to wear gloves all day. During their stay in kindergarten, children also perform numerous activities (changing clothes, putting on/taking off shoes, feeding, washing hands, and sleeping and resting). Some children wear clothes inside out to prevent skin contact with rough labels and seams. The way of dressing should be individual and adapted to the needs and wishes of each child. Footwear must be anatomical, soft and wider than the foot due to the padding. It is recommended to buy shoes with velcro straps, and it is necessary to encourage children to perform the mentioned activities independently [13]. When washing hands and showering, it is important to pay attention to the temperature of the water and the flow, which should be moderate. After washing, it is recommended not to wipe the hands and body, but pat them lightly with a towel [9]. Digestion and eating can be a big problem and challenge if the mucous membrane of the mouth and esophagus is affected. The appearance of sores in the mouth and throat is particularly painful and can cause an aversion to certain foods. Problems with chewing and swallowing food can affect the type and texture of food a child will consume. Some children will need support when cutting food or when using cutlery, so we can advise the use of cuffs on the utensils, anti-slip mats, etc. In addition to feeding, it is important to mention the drinking activity for which it is recommended to use straws and adapted glasses, for example glasses with handles [13]. Brushing teeth can be difficult due to microstomia [30], and OT intervention can be the use of a soft brush with short bristles. Going to the toilet can be made easier by adding a soft attachment to the toilet bowl that will make sitting and standing up easier and will not damage the child's skin. During sleeping activities, it is necessary to protect the edges of the bed with soft material and to make the necessary additional modifications related to the adaptation of the bed (additional pillows, soft sponge or pads) [9]. *OT intervention in the school period.* OT has an important role in ensuring a safe stay and quality integration of the child in school through the education of the professional team and children (to prevent peer violence and stigmatization), adaptation of the environment, drawing and writing accessories, counseling parents on the purchase of a school bag and possible adaptations that would facilitate the performance of daily activities. Acquiring a school bag is very important and choosing it is a joyful event for the first grader. It is recommended to get a light bag with soft backpacks and lining their edges or bags on wheelsPreparing and adjusting the environment includes protecting sharp edges with a soft sponge, arranging or removing unnecessary furniture in classrooms. The child must be provided with a place to sit that is away from radiators or windows so that they are not exposed to heat and sunlight, which can promote the formation of blisters [9]. Writing can be made easier through the adaptation of writing utensils, for example, silicone or rubber tips and thickenings on pens. Writing movements in children with EB come more from the wrist, elbow and shoulder and cause great fatigue. The participation of children with EB in the teaching of technical and artistic as well as physical and health education presents a special challenge and requires the most adjustments. When carrying out certain activities, it is important to pay attention to the material, which must not have sharp edges and a rough surface. It is extremely important to encourage the child to participate in sports and recreational activities. They will help the child develop gross motor skills, improve coordination and posture, help them gain various experiences and make them aware of their own limits with regard to the pain present. The goal of the OT intervention is to enable the child, despite the limitations, to experience everything that other children do and to achieve as much independence as possible in all aspects of school life. Recommendations for dressing are the same as in previous development periods, with education on certain ways of adjusting clothing items, for example, a thickened jacket zipper that would facilitate functional hand grip and hand movement when dressing and undressing. Patients are recommended to wear cotton and silk clothing, without rough labels, irregular seams or tight elastic. Also, it is recommended to wear sports clothes because they are breathable and prevent sweating, which normally leads to the formation of more blisters. Problems and challenges for feeding have been described previously, but it is important to note that parents will best inform the school about their child's nutritional needs and about adapted cutlery if their child uses it. *OT* *intervention in adolescence and adulthood.* Adolescence is a unique period, it marks the transition from childhood to adulthood. Adolescents want to achieve independence, fit into society and be accepted, adolescents with EB have the same wishes. In this period, the external appearance becomes very important, and at the same time, they are more and more prone to the formation of new wounds and blisters, which leads to visible scars. The OT can recommend establishing contacts with other adolescents with EB and provide support from community associations. For girls, the make-up activity is a priority, therefore RT recommends products with suitable ingredients that will not harm the skin and, if necessary, adjusts accessories for the make-up activity. It is important to encourage patients to participate in social activities in order to more easily express their own opinions, attitudes, emotions and establish new acquaintances. In this period, the question of a future career arises, so people with EB need to be advised about employment opportunities. When counseling OT, one should take into account the possibilities and limitations, as well as the interests and existing talents of the individual. Most adults with EB want their own apartment, job, partner and driver's license, which can be achieved if the person is motivated and has a good support system [28]. Over the years, adults with EB develop certain compensatory strategies, habits and routines, and then the intervention primarily involves ergonomic adaptation of aids and the environment with the aim of maintaining optimal functioning in everyday activities. OT advises patients on the acquisition and use of aids. Aids for dressing (aid for putting on socks or buttoning), preparing food (food holder, jar opener) and doing housework (sponge/dishwasher brush holder) can be very helpful. The adjustment of the feeding accessories is the same as in the previous stages. The ergonomic adjustment of the home includes toilet lifts, transfer boards, removal of carpets, fixing of wires and cables, etc. People with EB can drive a car, but for successful and safe driving, certain modifications are required, such as additional mirrors, adjustment of the steering wheel according to grips, automatic gearbox, soft seats, automatic door locking and padded seat belts.

Support system and intersectoral cooperation

Intersectoral cooperation is a very complex process and is a prerequisite for interdisciplinary cooperation. Different systems with their complexities and separate protocols participate in this process through a partnership relationship, sharing responsibilities and benefits. The early intervention model implies the connection of the health system, social care and the educational system. This form of cooperation has not fully taken root in Croatia. However, in Croatia, the association DEBRA, the Society of Epidermolysis Bullosa patients, which was founded in 1996 with the aim of connecting families of EB patients, stands out for its activities. DEBRA is a member of the international organization of patients with epidermolysis bullosa DEBRA International, the Croatian Association for Rare Diseases and the European Organization for Rare Diseases EURORDIS and is part of the International Alliance of Dermatology Patients Organizations (GlobalSkin). The activities of the association include the development of intersectoral cooperation for the improvement of health and social care and support for patients with EB and other rare dermatological diseases, psychological support, implementation of early intervention and occupational therapy for patients, development of mobile teams, advocacy and lobbying to improve the daily life of patients with rare diseases. Intersectoral cooperation should be aimed at improving the social and health care system, helping with integration and participation in the community, and advocating the rights of the sick.

Conclusion

EB is a rare dermatological disease that leads to a series of disabilities that limit the performance of daily occupations within the home and community and make it difficult for the sufferers to realize their personal goals. OT is a member of the interdisciplinary team and has an important role in the habilitation and rehabilitation of EB patients and their families. The goal of the OT process is to improve and increase the functional abilities and QoL of both patients and their families to experience numerous life situations despite the limitations. The OT process changes according to the age and needs of the patients and their families with frequent repeated evaluations, empowerment of parents, adaptation of the environment and implementation of training activities. The emphasis is also on an individual approach and intersectoral cooperation that would maximize integration and participation in the community and advocate the rights of patients.

References

[1] Fine J, Bruckner-Tuderman L, Eady R, Bauer E, Bauer J, Has C, et al. Inherited epidermolysis bullosa: updated recommendations on diagnosis and classification. J Am Acad Dermatol. 2014;70:1103–26.

[2] Debra Hrvatska: Društvo oboljelih od bulozne epidermolize. Posjećeno na: 26.03.2024. Dostupno na https://www.debra.hr/

[3] Dugonić M. Bulozna epidermoliza – uzrok, simptomi i liječenje. Kreni zdravo. Posjećeno na: 08.04.2024 Dostupno na:https://krenizdravo.dnevnik.hr/zdravlje/bolesti-zdravlje/bulozna-epidermoliza-uzrok-simptomi-i-lijecenje

[4] The WHOQOL Group. The World Health Organization quality of life assessment (WHOQOL): position paper from the World Health Organization. Social Science and Medicine.1995;41(10), 1403–1409.

[5] Both H, Essink-Bot ML, Busschbach J, Nijsten T. Critical review of generic and dermatology-specific health-related quality of life instruments. Journal of Investigative Dermatology. 2007;127(12), 2726–2739.

[6] Frew J, Martin L, Nijsten T, Murrell D. Quality of life evaluation in epidermolysis bullosa (EB) through the development of the QOLEB questionnaire: An EB-spe- cific quality of life instrument. Br J Dermatol. 2009;161(6):1323–30.

[7] Jain SV, Murrell DF . Psychosocial impact of inherited and autoimmune blistering diseases. Int J Womens Dermatol. 2018;4(1):49–53 .

[8] Haynes L. Nutritional support for children with epidermolysis bullosa. British Journal of Nursing. 2006; 15(20), 1097–1101.

[9] Fine JD, Bruckner-Tuderman L, Eady RA, Bauer EA, Bauer JW, Has C et al. Inherited epidermolysis bullosa: Updated recommendations on diagnosis and classification. Journal of American Academy of Dermatology. 2014; 70(6), 1103–1126.

[10] Caio Zidorio AP, Dutra ES, Dias Leao OD, Carvalho Costa IM. Nutritional aspects of children and adolescents with epidermolysis bullosa: Literature review. Anais Brasileiros de Dermatologia. 2015;90(2), 217–223.

[11] Sebaratnam DF, McMillan JR, Werth VP, Murrell D. Quality of life in patients with bullous dermatoses. Clinical Dermatology. 2012;30(1), 103–107.

[12] Debra Hrvatska. Integracija djece s buloznom epidermolizom (EB) u sustav predškolskog odgoja i obrazovanja. 2022. Posjećeno na: 31.03.2024. Dostupno na https://www.debra.hr/integracije-djece-s-buloznom-epidermolizom-u-sustav-predskolskog-odgoja-i-obrazovanja/

[13] Weiβ H, Prinz F. Occupational Therapy in Epidermolysis Bullosa. Beč: Springer-Verlag. 2013.

[14] Chan JM, Nicole A, Segovia A, Ladd L. Quality of Life and Independence in Activities of Daily Living in Epidermolysis Bullosa. Research Square. 2020. Posjećeno na: 26.03.2024. Dostupno na DOI: https://doi.org/10.21203/rs.3.rs-41738/v1

[15] Bartolac A. Procjena okupacija u radnoj terapiji. Zagreb: Zdravstveno Veleučilište, 2016.

[16] Pizzi MA, Richards LG. Guest Editorial—Promoting health, well-being, and quality of life in occupational therapy: A commitment to a paradigm shift for the next 100 years. American Journal of Occupational Therapy. 2017;71, 7104170010. Posjećeno na: 28.03.2024. Dostupno na https://doi.org/10.5014/ajot.2017.028456

[17] Bartolac A. Što je znanost o okupaciji? Istraživački pristup aktivnostima svakodnevnog života, 2013. Posjećeno na: 30.03.2024. Dostupno na: https://hrcak.srce.hr/file/184623

[18] Chan JM, Weisman A, King A, Maksomski S, Shotwell C, Bailie C, et al. Occupational therapy for epidermolysis bullosa: clinical practice guidelines. Orphanet Journal of Rare Diseases. 2019;14:129

[19] Tuntland H, Aaslund MK, Langeland E, Espehaug B, Kjeken I. Psychometric properties of the Canadian Occupational Performance Measure in home-dwelling older adults. J Multidiscip Healthc. 2016;9:411–423

[20] Langeland E, Tuntland H, Folkestad B, Forland O, Jacobsen FF, Kjeken I. A multicenter investigation of reablement in Norway: a clinical controlled trial. BMC Geriatrics. 2019;19:29.

[21] Öksüz Ç, Akel BS, Aran OT, Sığırtmaç İ, Leblebicioğlu AG. Do hand outcome measures reflect cultural influences? Acta Orthop Traumatol Turc. 2017;51:325–330.

[22] Tuntland H, Kjeken I, Folkestad B, Førland O, Langeland E. Everyday occupations prioritised by older adults participating in reablement. A cross-sectional study. Scand J Occup Ther. 2020;27:248–258.

[23] Enemark Larsen A, Rasmussen B, Christensen JR. Enhancing a client-centred practice with the Canadian Occupational Performance Measure. Occup Ther Int. 2018.

[24] Stevens A, Koke A, van der Weijden T, Beurskens A. The development of a patient-specific method for physiotherapy goal setting: a user-centered design. Disabil Rehabil. 2018;40:2048–2055.

[25] Flink M, Bertilsson AS, Johansson U, Guidetti S, Tham K, von Koch L. Training in client-centeredness enhances occupational therapist documentation on goal setting and

client participation in goal setting in the medical records of people with stroke. Clin Rehabil. 2016;30:1200–1210.
[26] Mahoney FI, Barthel DW. Barthel index. Maryland state medical journal. 1965.
[27] Kramer JM, Kielhofner G, Smith EV. Validity evidence for the Child Occupational Self Assessment. American Journal of Occupational Therapy. 2010;64(4), 621-632.
[28] Debra Hrvatska: Društvo oboljelih od bulozne epidermolize. Posjećeno na: 20.03.2024. Dostupno na https://www.debra.hr/wp-content/uploads/2020/11/Debra-brosura.pdf
[29] Great Ormond Street Hospital for Children, GOSH NHS Foundation Trust. The new baby with Epidermolysis Bullosa (EB): information for families. 2020. Posjećeno na: 14.04.2024. Dostupno na https://media.gosh.nhs.uk/documents/EB_new_baby_F2126_FINAL_Apr20.pdf
[30] Matijać T, Bulić D. Promicanje međusektorske suradnje u modelu rane intervencije. Paediatr Croat. 2021;65:174-9. Posjećeno na: 04.06.2023. Dostupno na: https://hrcak.srce.hr/file/405430.

RADNOTERAPIJSKI PROCES KOD BULOZNE EPIDERMOLIZE

Edina Pulić[1], Manuela Komorski[2], Jelena Klanjčić[3], Tatjana Njegovan Zvonarević[1], Maja Trulec[4]

[1]Zdravsteno veleučilište, Mlinarska cesta 38, Zagreb, Republika Hrvatska
[2]Hrvatski savez gluhoslijepih osoba Dodir, Medulićeva 34, Zagreb, Republika Hrvatska
[3]Specijalna bolnica za kronične bolesti dječje dobi Gornja Bistra, bolnička ulica 21, Gornja Bistra, Republika Hrvatska
[4]Debra, Društvo oboljelih od bulozne epidermolize, Prilaz Gjure Deželića 30, Zagreb, Republika Hrvatska

Sažetak. *Bulozna epidermoliza (EB) pripada skupini rijetkih, genetskih bolesti za koje je karakteristična izuzetna osjetljivost kože, a u nekih oblika i sluznice. Progresivna priroda ove bolesti narušava život oboljele osobe te postupno dovodi i do invaliditeta. To je nasljedna bolest koja uzrokuje jaku osjetljivost kože. Na najmanji pritisak ili trenje može doći do pojave bolnih mjehura i rana, a komplikacije mogu nastati i na sluznicama (oči, usta, probavni sustav). U radu je prikazan utjecaj EB na kvalitetu života, povezanost okupacija i zdravlja, radno terapijski (RT) proces kod EB, sustav podrške i međusektorska suradnja. Naglasak je stavljen na progresiju, problematiku i utjecaj bolesti na svakodnevni život oboljelih, od rođenja do prelaska u odraslu dob. Sukladno tome prikazane su RT intervencije, čiji je primarni cilj prevladavanje prepreka i osnaživanje okupacijske participacije u svrhu unaprjeđenja kvalitete života oboljelih i njihove participacije u aktivnostima svakodnevnog života. Rad pruža uvid u kompleksnost ovog područja, uzimajući u obzir istraživanja na području RT prakse i kvalitete života oboljelih od EB.*

Ključne riječi: *bulozna epidermoliza, radno terapijski proces, kvaliteta života, okupacijska participacija*