

The selected health problems in children with Stevens–Johnson syndrome: Nursing care concept

Wybrane problemy zdrowotne dziecka z zespołem Stevensa i Johnsona – koncepcja opieki pielęgniarskiej

Grażyna Cepuch^{A–F}, Agnieszka Kruszecka-Krówka^{B–D}

Faculty of Health Sciences, Jagiellonian University Medical College, Cracow, Poland

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 the article

Pielęgniarstwo i Zdrowie Publiczne, ISSN 2082-9876 (print), ISSN 2451-1870 (online)

Piel Zdr Publ. 2019;9(4):301–308

Address for correspondence

Agnieszka Kruszecka-Krówka
E-mail: agnieszka.kruszecka-krowka@uj.edu.pl

Funding sources

None declared

Conflict of interest

None declared

Received on March 19, 2019

Reviewed on July 9, 2019

Accepted on September 9, 2019

Abstract

Stevens–Johnson syndrome (SJS) is a life-threatening disease affecting the skin and mucous membranes, frequently causing systemic complications, most often occurring as an undesirable reaction to pharmacological treatment. Because of the widespread use of polypharmacotherapy, the incidence of the syndrome increases, also in the pediatric group. The skin is recognized as so-called signal organ and all kinds of changes occurring on it are an ideal indicator helpful in the diagnosis of adverse drug reactions. The aim of the article is to present the health problems in children with SJS and to show how these problems can be solved thanks to the actions undertaken by the nursing team. A child with SJS should be treated by qualified staff in the intensive care unit or a burns ward. Skin and mucosal changes cause high-intensity pain and can be a source of systemic and local infection. Constant supervision of the patient, vital signs monitoring, assessment of changes on the skin and mucous membranes, their proper management, as well as the participation in the treatment are the basis for nursing activities.

Key words: drug side effects, child, health problems, Stevens–Johnson syndrome

This is a translated article.

Please cite the original

Polish-language version as

Cepuch G, Kruszecka-Krówka A. Wybrane problemy zdrowotne dziecka z zespołem Stevensa i Johnsona – koncepcja opieki pielęgniarskiej. *Piel Zdr Publ.* 2019;9(4):301–308. doi:10.17219/pzp/112221

DOI

10.17219/pzp/112221

Copyright

© 2019 by Wrocław Medical University

This is an article distributed under the terms of the Creative Commons Attribution 3.0 Unported License (<https://creativecommons.org/licenses/by/3.0/>)

Streszczenie

Zespół Stevensa i Johnsona jest schorzeniem zagrażającym życiu pacjenta, dotyczącym skóry i błon śluzowych, z częstymi powikłaniami układowymi, występującym najczęściej jako niepożądana reakcja na zastosowane leczenie farmakologiczne. Ze względu na szerokie stosowanie polifarmakoterapii wzrasta prawdopodobieństwo rozwinięcia się zespołu również w grupie pacjentów pediatrycznych. Skóra uznawana jest za tzw. narząd sygnałowy, dlatego wszelkiego rodzaju zmiany na niej występujące są idealnym wskaźnikiem pomocnym w diagnostyce niepożądanych reakcji polekowych. Celem artykułu jest przedstawienie problemów zdrowotnych dziecka, u którego rozpoznano zespół Stevensa i Johnsona, oraz zaproponowanie metod ich rozwiązania w działaniach zespołu pielęgniarstwa. Dziecko zmagające się z zespołem Stevensa i Johnsona powinno być leczone przez wykwalifikowany personel na oddziale intensywnej terapii lub oddziale zajmującym się leczeniem ran oparzeniowych. Zmiany skórne i śluzówkowe powodują ból o dużym natężeniu i mogą być źródłem zakażenia ogólnoustrojowego i miejscowego. Stały nadzór nad pacjentem, monitorowanie parametrów życiowych, ocena zmian stanu skóry i błon śluzowych oraz ich pielęgnacja, a także udział w leczeniu stanowią podstawę opieki pielęgniarstwa.

Słowa kluczowe: działania niepożądane leków, dziecko, problemy zdrowotne, zespół Stevensa i Johnsona

Introduction

Stevens–Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are considered multi-organ syndromes endangering the life and health of the patient. They constitute a mucocutaneous reaction connected with hypersensitivity to medications¹ or, decidedly more sporadic, an infectious agent. The course of SJS and TEN often leads also to system complications.^{2,3}

The risk of development of SJS in the pediatric patient group is connected with the application of mainly sulfonamides (i.a., sulfasazaline, co-trimoxazole), penicillin, non-steroid anti-inflammatory medication, cephalosporin, quinolones, iodine contrast agents, xenogeneic serums, and anti-seizure medication (i.a., lamotrigine, phenytoin, carbamazepine, phenobarbital) during the therapy,^{4–6} and medication applied in cancer treatment.⁷ Above all else, *Mycoplasma pneumoniae* is listed among infectious agents connected with the emergence of SJS.^{8,9} However, there is still no clear answer whether the infectious agent is only a cofactor or the trigger agent in connection with the medication, and whether it may be directly responsible for causing the SJS.¹⁰

It is deemed that there are 2 groups of risk factors for the development of SJS. The 1st group consists of disease factors such as: viral infections, chronic inflammatory diseases or diseases reducing immunity (e.g., human immunodeficiency virus (HIV) and systemic lupus erythematosus),^{11–13} while the 2nd group consists of genetic predispositions.^{14–16} Alcohol is also considered to be a probable risk factor for the development of SJS.^{17,18} The latter cannot be ruled out in pediatric patients group, mainly adolescents.

The exact pathomechanism of SJS development has not been fully discovered. The disease is probably the consequence of many characteristic allergic-immunologic cellular and humoral reactions.^{15,19} The damage done to the metabolic pathway of the medication taken is considered to be the main reason for the emergence of SJS. It is then that the accumulation of its toxic metabolites takes place

within the organism. Toxins, viruses or immunological mechanisms can be the cause of intensified apoptosis of keratinocytes taking place in SJS. Another causal agent may be programmed death surface receptors of a cell.^{16,20}

Clinical symptoms

Prodromes, such as: headache, fever, sore throat, dysphagia, pruritus, and eye irritation precede the emergence of the actual symptoms of the disease by 1–10 days. Typical SJS symptoms are characterized by the creation of petechiae, blisters and erosions of oral mucosae, genitourinary organs, upper section of the digestive tract, or upper respiratory tract. The infiltration of oral mucosae is observed in 90% of SJS cases.²¹ Skin changes may take the form of erythematous skin lesions of any part of the body. Hemorrhagic erosions frequently emerge on the vermilion zone, which dries, creating layers of scabs. Meanwhile, the inflammatory state of mucosa of the esophagus leads to the emergence of dysphagia and odynophagia. Many patients experience also conjunctivitis and keratitis. Pathological changes in the oculus are determined in approx. 75% of patients.²² The characteristic feature of SJS is its abrupt commencement, high body temperature, joint and muscle pain, and dysuria.^{2,6,23,24}

In severe cases of SJS, pneumonia, myocarditis, nephritis, hepatitis, and secondary skin infections may take place, as well as heavy immune-mediated kidney and liver damage. The patient is also threatened by the emergence of meningitis or sepsis due to the general inflammatory reaction of the organism to the infection.²⁴

Distant complications may also take place during the course of SJS. They pertain, above all else, to respiratory, digestive and genitourinary system, and are the result of the proceeding scarring of mucosae, resulting in the disruption of food intake, obturation of the respiratory tract and symptoms of dysuria. Patients suffering from SJS are threatened by complications of the oculus,²⁵ such as: xerophthalmia, photophobia, chronic conjunctivi-

tis, endophthalmitis, fusion of the eyelid with the eyeball, creation of scars in the area of conjunctiva, cornea softening, and excessive accretion of keratin in the corneal epithelium, which may lead to blindness.²³ Late SJS complications include skin discoloration, fingernail deformation and anthropic changes of nail plates, chronic ulcerations of the oral cavity and reproductive organs, and atrophy of tongue papillae. The emergence of late complications is observed in 47% of patients.²⁶ Stevens–Johnson syndrome relapse is generally of more serious course than the first episode of the disease and may end in extreme cases with the patient's death, especially in the event of 30% of the patient's body surface area being taken over by it and the development of toxic epidermal necrolysis.^{16,21}

Treatment

There are no recommendations as to the treatment of SJS, also for the group of children and teenagers, and the recommended plans of proceeding are determined on the basis of in vitro examinations and descriptions of a series of cases. The basis of the proceeding is a quick diagnosis of the disease, elimination of the factor causing the reaction and implementation of treatment as soon as possible.^{6,27} Medications of symptomatic action, both local as well as general, are applied in the treatment of SJS. Local therapy consists in the application of antifungal and antiviral medication, antibiotics, glucocorticosteroids and anesthetics onto the skin and mucosae, and, in the event of such recommendation, the implementation of the so-called moist wound healing with the use of modern bandages. In the case of deep or broad skin changes, surgical wound cleaning and covering them with biological bandages, biosynthetics or autologous skin transplants are applied, and the therapy takes place in isolation.³ Antibiotics, antiviral medication and antihistamines, immunoglobulins and/or glucocorticosteroids, as well as diuretics are used during the treatment. Tetanus preventive care implementation is also recommended.²⁸ Second-choice methods and medications are: plasmapheresis,²⁹ infliximab or cyclosporine. This includes also hemodiafiltration in the event of developing kidney failure.³⁰ In the case of the patient's worsening state in the course of SJS or the development of toxic epidermal necrolysis, modification and intensification of the applied treatment is necessary.^{30,31} However, the effectiveness of the therapy based upon the application of glucocorticosteroids, immunoglobulins, cyclosporines, biological medication, or plasmapheresis is uncertain,^{3,32} and in the case of glucocorticosteroids – even controversial.^{33,34} There is no unequivocal position on the preventive antibiotic therapy intake.

Hospitalization of patients afflicted with SJS should take place in the burns treatment unit or intensive care

unit with the participation of a multidisciplinary team (dermatologist, dentist, plastic surgeon, ophthalmologist, pulmonologist, gastrologist, urologist, physiotherapist) and with the inclusion of meticulous care in terms of wounds, hydration, pain control, and isolation from other patients.^{6,28,35} Provision of continuous nursing care is of key importance for the success of the therapy. Good cooperation of the entire treatment team allows to achieve positive therapy results or to avoid or minimize the consequences of complications.³⁶

Diagnostic and treatment & nursing tasks of the nursing team

1. Elimination of pain connected with pathological changes of the skin and mucosae:

- regular pain assessment of the pain: qualitative (pain characteristic) and quantitative (pain intensity degree with the use of, e.g., visual analog scale (VAS) and numeric rating scale (NRS)),³⁷ the time of its duration and location and the assessment of the effectiveness of the undertaken analgesic actions documented in the pain assessment sheet, allowing for the effective adjustment of the analgesic proceeding to the needs of the child with the inclusion of the current need for painkillers (e.g., prior to bandage change, prior to diagnostic examination);
- assessment of the emotional state and behavior of the patient – depressed mood and emotional state featuring characteristics of depression and anxiety lower the pain threshold and increase the need for painkillers. It is advisable to assess the entirety of the pain management procedure in the form of a protocol;
- assessment of the state of skin and oral mucosae, oculus and crotch area in terms of the intensification/recession of changes allows for the assessment of the effectiveness of the undertaken basic treatment and modification of the applied analgesic treatment;
- participation in the basic treatment of the disease in accordance with the individual patient's treatment record:
 - participation in the intake of medication of systematic action (immunoglobulin, glucocorticosteroids, antibiotics, metronidazole, immunosuppressive medications). During the administration of medication of systematic action, it is advisable to perform a thorough assessment of the patient's state due to the possibility of disease process exacerbation. Basic treatment of the disease reduces the amount of changes and, as a result, sources of pain sensations are eliminated³⁸;
 - participation in local treatment of skin changes – the following are applied (i.a.): neutral paraffin gases, enzymatic bandages, polyurethane bandages, hydrogel, hydrofiber, hydrocolloid bandages, antibiotic,

glucocorticosteroid or antifungal medicinal preparations, bandages with silver ions, and specialist procedures with the application of silicone.³⁹ Treatment of skin changes eliminates main pain sources. Application of modern bandages during therapy reduces the pain experienced during their change since, thanks to the gelatinization process during the contact with the exudation, the bandages do not adhere to the tissues. Nevertheless, it is important to lubricate the skin during the period of wound healing and scar emergence. Furthermore, one should absolutely exclude the application of traditional skin bandages, the removal of which may cause pain and skin injuries connected with the application of medical adhesives (medical adhesive-related skin injuries (MARSII)), as well as irritating solvents for patch removal (i.e., pharmaceutical petrol). One should use hypoallergenic, non-alcoholic products on the basis of mineral oil or silicone or water-soaked gauze pads for the removal of medical adhesives (patches) by constantly moisturizing the surface at the border of the adhesive and skin⁴⁰;

- participation in the local treatment of ocular mucosa – in this case, anti-infective, anti-allergic and anti-inflammatory medications, as well as medications which improve metabolism and cellular regeneration, and moisturizing medications in the form of drops, creams and gels. Preservative-free medications are recommended^{24,41};
- participation in local treatment of ulcerations of oral mucosa – treatment of local changes in the oral cavity is symptomatic and depends on the degree of their intensification.³⁹ The purpose of the oral cavity care in the patient who shows signs of ulcerations of the mucosa during the course of SJS, is, above all else, the maintenance of the oral cavity in a state of cleanliness (removal of food remains and dead cells from the oral cavity), dental caries prevention, elimination of ulcerations, and prevention of the creation of new disease changes on the mucosa, as well as providing comfort to the patient. Antiseptic medications (antibacterial washes, e.g., Octenidol, and local antifungal medications, e.g., nystatin) are used during the treatment of inflammatory changes of oral cavity mucous membranes, as well as an anesthetic ones (e.g., ones containing lignocaine, benzocaine). One can sprinkle the oral cavity with saline solution or wash it with aqueous solution of gentian or benzydamine. Preparations accelerating epithelialization and protecting the mucous membrane against mechanical damage (e.g., washing of the oral cavity with linseed infusion) are also used in local treatment of disease changes within the oral cavity.³⁹ If the patient can consume meals, anesthesia of the mucous membrane needs to be performed 10–15 min prior to the planned meal and the intake of food and

drinks, which irritate disease changes and cause pain (hard, spicy, sour, hot), is to be avoided. Sterile gauze pads and saline solution need to be provided for oral cavity care and during the execution of hygienic procedures one needs to avoid rubbing the mucous membrane in order to reduce the risk of secondary damage creation. It is necessary to provide constant lubrication of the vermilion zone with hypoallergenic lubricating or moisturizing preparations, e.g., containing vitamin A or ones such Be-panthen.²¹ All diagnostic, therapeutic and nursing procedures are to be executed in a way which does not intensify pain.

The basic goal of an effective analgesic action is to provide the pediatric patient with subjective comfort and to stop or not allow for occurrence of a cascade of pathophysiological processes of the organism, which could be an effect of an ineffective pain treatment. When planning care, one needs to take into account also the application of non-pharmacological methods of pain alleviation and to prepare the parents to participate in care at each stage of treatment.

2. Reduction of the risk of wound infection or development of sepsis due to skin blisters rupturing and damage of the mucous membrane, pharmacological treatment (antibiotics, glucocorticosteroids, immunosuppressive medications) and immunity disruptions during the course of SJS:

- assessment of the current state of the child in regular time intervals resulting from the current health status, including:
 - measurement of vital signs (heart rate, blood pressure, body temperature) according to the indications resulting from the current health status. Body temperature increase and tachycardia may indicate the development of the infection. However, one should remember that the increase of temperature takes place also due to intensified catabolic processes taking place within the organism, inflammatory response of the organism or pain accompanying the disease. While administrating antipyretic medications, one must maintain caution and avoid applying medications which could become the cause of SJS symptoms and duplicating the intake of medications from the same group, e.g., non-steroid anti-inflammatory medications¹⁰;
 - assessment of the patients behavior in order to detect abnormalities (e.g., apathetic behavior, excitation, disturbance of consciousness) constituting proof of infection development;
 - assessment of the state of skin and mucous membranes in terms of the emergence of wound infection development features (assessment of the local infection pattern – skin change color and possible wound exudation, scent, pain intensification, tissue swelling,

local warming) or hemorrhagic petechiae, which may constitute the proof of sepsis development⁴¹;

- collection of swabs from disease-changed locations, provided that it is recommended. Patients afflicted with SJS carry the risk of secondary infections of skin and mucous membranes with pathogenic microorganisms, such as bacteria (e.g., staphylococci, streptococci, *Clostridium*) or fungi. The biological material is to be collected strictly according to the recommendations of the laboratory and it is to be delivered to the lab as quickly as possible;
- intake of medications of systematic and local action (see item 1). During the period of empirical or targeted antibiotic therapy, one must maintain caution due to the risk of exacerbating the disease process in response to the applied treatment, which can be confirmed with, i.e., newly emergent changes or the worsening of the already existing skin and mucous membrane changes;
- taking baths in potassium permanganate, use of sterile bed linen³⁹ and change of personal and bed underwear at least once per day and according to recommendations;
- selection of optimal modern bandages adapted to the patient's needs, type of wound and healing stage, which maintain the moist environment of the wound, absorb the possible wound exudation, provide quick and effective antibacterial activity as needed, and enable gas exchange while favoring protection of the skin surrounding the wound against maceration and secondary damage while changing the bandage. Absorption and durability of bandages made of alginates, hydrogels, hydrofibers, and hydrocolloids make it possible for the bandage to remain on the wound for a longer period of time. These bandages close the exudation and harmful components within their structure, thanks to which they prevent cross-infections. Bandages, the contact layer of which contains silver ions, can be used for covering the infected wounds or ones suspected of being infected and ensure effective combating of a wide range of microorganisms, including methicillin-resistant *Staphylococcus aureus* (MRCA), vancomycin-resistant *Staphylococcus aureus* (VRCA) and other antibiotic-resistant strains.⁴² Regarding patients afflicted with SJS, the wound treatment protocols are somewhat similar to burn treatment protocols and are to be strictly followed;
- taking into account the subjective feelings of the patient when planning care – subjective symptoms may precede objective symptoms. Reaction to the patient's remarks may enable the undertaking of early intervention by the therapeutic team;
- thorough assessment of vessel catheter locations – these locations constitute often sources of infection, especially in patients of weakened immunity. It is beneficial to apply semipermeable membranes featuring high moisture vapor transmission rate (MVTR) in order to secure peripheral and central vessel catheter locations,⁴³ which enable their assessment in terms of the presence

of features of local inflammatory state, constitute protection against mechanical damage and soiling, constitute a barrier for bacteria and other microorganisms, and allow for washing the contaminated area of the body which is, e.g., exposed to contact with excrements. This bandage does not require to be changed on a daily basis;

- caring for crotch and urethra vicinity – it is necessary to maintain cleanliness of the crotch and buttocks area by daily precise toilet with gentle cleaning products (also every time following urine or stool being expelled) and gentle drying. It is recommended to apply disinfection preparations (e.g., potassium permanganate solution, octenidine-based solution) and medicinal ones (e.g., preparations with chloramphenicol, mupirocine or detreomicin, fluticasone propionate or betamethasone valerate) in some patients⁴¹ for skin changes in crotch vicinity. The disease process in this location breaches the natural protective barrier of the urinary tract and may favor the development of inflammatory state of the urinary system. When protecting the urinary tracts against infection, it is important to hydrate the patients and encourage them to expel the urine. If the patient has the suprapubic catheter, one needs to take care for the catheter not to rub the skin, since it increases the risk of intensifying the changes and their infection. The delicate skin around the wounds can be protected against harmful effect of dampness, mechanical irritation or soiling by using the so-called liquid barrier membranes: foams, tissues or sprays, which may contain silicones, acrylates, organic polymers or non-organic compounds dispersed within the solvent. Barrier products reduce the risk of MARSII emergence by smoothing the skin and creating a thin, microscopic protective barrier⁴⁰;
- intake of medications into the conjunctival sac in strict accordance with assumed procedures – diligent assessment in terms of the emergence of new erosions or croupous membranes is applicable. Only sterile gauze pads can be applied when caring for eyes and when performing medical actions;
- provision of frequent variation of the patient's position in bed in order to reduce pressure and ischemia in the area of locations changed by the disease (every 1 h or 2 h, depending on the state of the skin and the patient's general status);
- meticulous adherence of the medical personnel to the application of personal protection equipment (such as: gloves, additional protective apron, facial masks) and to the rules of asepsis and antiseptics;
- preparations of parents to caring for the child, including training within the scope of the rules of sanitary regime maintenance and caring for the afflicted child;
- preparation of the patient to the procedure of bandage replacement while under general anesthesia according to the individual prescription sheet.

3. Maintenance of hydro-electrolyte balance of the child and provision of nutrients according to requisition

Fever, damaged skin, changes on oral cavity mucous membranes, or impeded intake of meals resulting from disease-related changes located within the digestive tract (erosions and scarring) favor disruptions in the organism's water balance and malnutrition. Possible hemorrhages from the digestive tract and disease-related changes, which may affect bone marrow, liver and pancreas, can favor anemia and malnutrition. Nursing actions should include:

- assessment of the child's hydration degree: body mass measurement, assessment of moisture content of oral cavity mucous membranes, skin flexibility, heart rate, blood pressure, capillary return, amounts of liquids taken in and expelled (liquid balance with the inclusion of possible additional liquid losses in the event of fever, emesis or gastroparesis). However, assessment of the skin and mucous membranes in terms of hydration of the child may be difficult due to the presence of disease changes;
- participation in diagnostic examinations according to the individual prescription sheet – it is of significance to prepare the patient psychologically for examination in order to reduce fear. Collection of arterialized blood for laboratory examinations, e.g., gasometric analysis of blood and ion levels, and morphotic examination of blood, requires the puncture location to be warmed up and to avoid blood extrusion due to the risk of examination result becoming altered (exclusion of pre-lab errors). Collection of venous blood for biochemical tests (e.g., in order to specify the concentration of total proteins and albumins in the serum) will also be required – the assessment of protein concentration is necessary in order to treat SJS-afflicted patient effectively; assessment of specific gravity of urine and the presence of ketone bodies allows for the monitoring of hydration state;
- taking care for the patient to feel as low level of stress during lab examination as possible;
- oral rehydration in accordance with daily water demand of the patient (taking into account their body mass). In the event of the lack of effectiveness of oral rehydration, rehydration via nasal-gastro tube and, in the event of lack of tolerance or the impossibility of enteral rehydration, infusion therapy according to the individual prescription sheet (coverage of liquid demand executed primarily by multi-electrolyte liquid intake). There may be a necessity to rehydrate according to the rules applied for a burn disease⁴⁴;
- intake of painkillers of systematic action or/and acting on oral cavity mucous membrane, according to the individual prescription sheet prior to the meal;
- provision of proper child nutrition – meal consistency adjusted to the child's capabilities (in hard cases: liquid diet in the beginning of the disease process, and follo-

wing that blended and light meals, prepared taking into account of the high-protein diet).^{8,11} In the event of the lack of possibility of implementing/maintaining oral feeding, it is necessary to feed via nasal-gastro tube or to apply parenteral nutrition according to the individual prescription sheet in order to provide the essential nutrition, necessary for wounds to heal. When placing the nasal-gastro tube, one must maintain extreme caution due to the possibility of hemorrhaging coming from the disease-changed mucous membrane of the esophagus;

- undertaking of the intervention to maintain/return the patient's thermal comfort (patients afflicted with SJS are at risk of pain, fever or excessive cooling). Therefore, maintenance of ambient temperature above 25°C, 30–32°C at optimum, is very important.³⁹ Disease process, fever, pain, and cooling of the organism are factors increasing protein catabolism within the organism, and they increase energy demand.

4. Maintenance of respiratory system performance

The disease process may also take over mucous membranes of the respiratory system, leading to the tightening of the respiratory system and pneumonia, which may result in the emergence of acute respiratory distress syndrome. Nursing care covers:

- assessment of airway, hemodynamic stability and general hydration state²⁸;
- diligent assessment of the functioning of the respiratory system (quantity and type of breath, child behavior, respiratory muscle work, pattern of breathing, skin coloration, auscultation of the thorax) according to the indications resulting from the current health status;
- constant monitoring via indirect patient observation or cardiometer with simultaneous interpretation of measurement results and periodic documentation in the case report form;
- reacting to the patient's remarks and their subjective feelings;
- encouraging to perform breathing exercises in order to reduce the risk of emergence of breathing disruptions resulting from the limitation of the patient's mobility, e.g., deepened inhalations and exhalations, breathing reinforced by arm movement, exhalation with resistance with the use of professional (e.g., Magic Ball apparatus with styrofoam/polystyrene ball, Triflo apparatus, Spiroflo apparatus) or unprofessional equipment (cotton pads, feathers, whistles, etc.);
- execution of nebulization in order to moisten the respiratory tract (e.g., 0.9% NaCl or 0.9% NaCl with hyaluronic acid);
- delicate care of the upper respiratory tract (shallow secretion extraction);
- airborne or systematic intake of medication according to the individual prescription sheet;
- on-demand intake of oxygen via oxygen mask, provided that it is recommended, or participation in passive

oxygen therapy according to the individual prescription sheet;

- provision of access to the equipment for resuscitation and intubation in the event of total tightening of the respiratory tract.

Professional and specialist nursing care for the SJS-afflicted child covers also the provision of support to the patient and their family, including the easing of the fear associated with the lack of self-acceptance. It is no less important to prepare the parents to care for the afflicted child with the maintenance of sanitary regime and particular care for its safety. It is of particular significance in the context of the planned discharge from the hospital due to the necessity of continuing skin and mucous membranes treatment commenced during the hospitalization and prevention of distant complication, including the emergence of hypertrophied scars.

Conclusions

Patients with SJS require holistic care of an interdisciplinary therapeutic team, including professional nursing care. Constant supervision of the patient, vital signs monitoring, assessment of changes on the skin and mucous membranes and their management, intake of medications of systematic and local action, as well as participation in nutritional therapy constitute the basis of nursing activities.

ORCID iDs

Grażyna Cepuch  <https://orcid.org/0000-0002-7124-565X>

Agnieszka Kruszecka-Krówka  <https://orcid.org/0000-0003-3974-6427>

References

- Sassolas B, Haddad C, Mockenhaupt M, et al. ALDEN, an algorithm for assessment of drug causality in Stevens–Johnson syndrome and toxic epidermal necrolysis: Comparison with case-control analysis. *Clin Pharmacol Ther.* 2010;88(1):60–68. doi:10.1038/clpt.2009.252
- Dodiuk-Gad RP, Chung WH, Valeyrie-Allanore L, Shear NH. Stevens–Johnson syndrome and toxic epidermal necrolysis: An update. *Am J Clin Dermatol.* 2015;16(6):475–493. doi:10.1007/s40257-015-0158-0
- Schwartz RA, Mc Donough PH, Lee BW. Toxic epidermal necrolysis: Part I. Introduction, history, classification, clinical features, systematic manifestations, etiology, and immunopathogenesis. *J Am Acad Dermatol.* 2013;69(2):173.e1–13. doi:10.1016/j.jaad.2013.05.003
- Levi N, Bastuji-Garin S, Mockenhaupt M, et al. Medications as risk factors of Stevens–Johnson syndrome and toxic epidermal necrolysis in children: A pooled analysis. *Pediatrics.* 2009;123(2):e297–304. doi:10.1542/peds.2008-1923
- Miliszewski MA, Mark G, Kirchhof MG, Sikora S. Stevens–Johnson syndrome and toxic epidermal necrolysis: An analysis of triggers and implications for improving prevention. *Am J Med.* 2016;129(11):1221–1225. doi:10.1016/j.amjmed.2016.03.022
- Chatproedprai S, Wuttichareonwong V, Tempark T, Wanankul S. Clinical features and treatment outcomes among children with Stevens–Johnson syndrome and toxic epidermal necrolysis: A 20-year study in a tertiary referral hospital. *Dermatol Res Pract.* 2018;2018:3061084. doi:10.1155/2018/3061084
- Kowalska M, Kowalik A. Dermatologic adverse events associated with chemotherapy and targeted anticancer therapy. *Przegl Dermatol.* 2016;103(2):127–138. doi:10.5114/dr.2016.59135
- Olson D, Watkins LK, Demirjian A, et al. Outbreak of mycoplasma pneumoniae: Associated Stevens–Johnson syndrome. *Pediatrics.* 2015;136(2):e386–394. doi:10.1542/peds.2015-0278
- Blanco N, Gutiérrez B, Valls I, et al. Stevens–Johnson syndrome in childhood. *Arch Soc Esp Otolmol.* 2017;92(5):241–244. doi:10.1016/j.oftal.2016.10.003
- Hinc-Kasprzyk J, Polak-Krzemińska A, Ożóg-Zabolska I. Toxic epidermal necrolysis. *Anaesthesiol Intensive Ther.* 2015;47(3):257–262. doi:10.5603/AIT.2015.0037
- Heelan K, Shear NH. Cutaneous drug reactions in children: An update. *Paediatr Drugs.* 2013;15(6):493–503. doi: 10.1007/s40272-013-0039-z
- Pavlos R, Mallal S, Ostrov D, Pompeu Y, Phillips E. Fever, rash, and systematic symptoms: Understanding the role of virus and HLA in severe cutaneous drug allergy. *J Allergy Clin Immunol Pract.* 2014;2(1):21–33. doi:10.1016/j.jaip.2013.11.005
- Yu J, Brandling-Bennett H, Co DO, Nocton JJ, Stevens AM, Chiu YE. Toxic epidermal necrolysis-like cutaneous lupus in pediatric patients: A case series and review. *Pediatrics.* 2016;137(6):e20154497. doi:10.1542/peds.2015-4497
- Ueta M, Tokunaga K, Sotozono C, et al. HLA class I and II gene polymorphisms in Stevens–Johnson syndrome with ocular complications in Japanese. *Mol Vis.* 2008;14:550–555.
- Chung WH, Hung SI. Recent advances in the genetics and immunology of Stevens–Johnson syndrome and toxic epidermal necrosis. *J Dermatol Sci.* 2012;66(3):190–196. doi:10.1016/j.jdermsci.2012.04.002
- Adler NR, Aung AK, Ergen EN, Trubiano J, Goh MSY, Phillips EJ. Recent advances in the understanding of severe cutaneous adverse reactions. *Br J Dermatol.* 2017;177(5):1234–1247. doi:10.1111/bjd.15423
- Łoboda J, Dudzik A, Chomyszyn-Gajewska M. Zespół Stevensa–Johnsona oraz zespół toksycznej nekrozy naskórka – na podstawie piśmiennictwa. *Przegl Lek.* 2015;72(1):35–37. http://www.wple.net/plek/numery_2015/numer-1-2015/35-37.pdf. Accessed on November 6, 2019.
- Bertulyte I, Schwan S, Hallberg P. Identification of risk factors for carbamazepine-induced serious mucocutaneous adverse reactions: A case-control study using data from spontaneous adverse drug reaction reports. *J Pharmacol Pharmacother.* 2014;5(2):100–138. doi:10.4103/0976-500X.130051
- Jung A, Kuligowski R, Kalicki B. Zespół Stevensa–Johnsona – obserwacje kliniczne. *Alergia.* 2005;3(25):42–44.
- White KD, Chung WH, Hung SI, Mallal S, Phillips EJ. Evolving models of the immunopathogenesis of T cell-mediated drug allergy: The role of host, pathogens, and drug response. *J Allergy Clin Immunol.* 2015;136(2):219–234. doi:10.1016/j.jaci.2015.05.050
- Creamer D, Walsh SA, Dziwulski P, et al. UK guidelines for the management of Stevens–Johnson syndrome/toxic epidermal necrolysis in adults 2016. *Br J Dermatol.* 2016;174(6):1194–1227. doi:10.1016/j.bjps.2016.04.018
- Guedry J, Roujeau JC, Binaghi M, Soubrane G, Muraine M. Risk factors for the development of ocular complications of Stevens–Johnson syndrome and toxic epidermal necrolysis. *Arch Dermatol.* 2009;145(2):157–162. doi:10.1001/archdermatol.2009.540
- Morales ME, Purdue GF, Verity SM, Arnoldo BD, Blomquist PH. Ophthalmic manifestations of Stevens–Johnson syndrome and toxic epidermal necrolysis and relation to SCORTEN. *Am J Ophthalmol.* 2010;150(4):505–510. doi:10.1016/j.ajo.2010.04.026
- Alerhand S, Cassella C, Koyfman A. Stevens–Johnson syndrome and toxic epidermal necrolysis in the pediatric population: A review. *Pediatr Emerg Care.* 2016;32(7):472–478. doi:10.1097/PEC.0000000000000840
- Catt CJ, Hamilton GM, Fish J, Mireskandari K, Ali A. Ocular manifestations of Stevens–Johnson syndrome and toxic epidermal necrolysis in children. *Am J Ophthalmol.* 2016;166:68–75. doi:10.1016/j.ajo.2016.03.020
- Finkelstein Y, Soon GS, Acuna P, et al. Recurrence and outcomes of Stevens–Johnson syndrome and toxic epidermal necrolysis in children. *Pediatrics.* 2011;128(4):723–728. doi:10.1542/peds.2010-3322
- Scott-Lang V, Tidam M, McKay D. Toxic epidermal necrolysis in children successfully treated with infliximab. *Pediatr Dermatol.* 2014;31(4):532–534. doi:10.1111/pde.12029

28. Lerch M, Mainetti C, Terziroli Beretta-Piccoli B, Harr T. Current perspectives on Stevens–Johnson syndrome and toxic epidermal necrolysis. *Clin Rev Allergy Immunol*. 2018;54(1):147–176. doi:10.1007/s12016-017-8654-z
29. Aihara Y, Oyama Y, Ichikawa K, et al. Toxic epidermal necrolysis in 4-year old boy successfully treated with plasma exchange in combination with methylprednisolone and iv immunoglobulin. *J Dermatol*. 2012;39(11):951–952. doi:10.1111/j.1346–8138.2012.01534.x
30. Hinc-Kasprzyk J, Polak-Krzemińska A, Głowacka M, Ożóg-Zabolska I. Zastosowanie plazmaferezy w zespole toksycznej nekrolizy naskórka u 4-letniego chłopca. *Anest Int Ter*. 2015;47(3):217–220.
31. Renke J, Sobkowiak M, Wołowska E. Przypadki poważnych działań niepożądanych po zastosowaniu farmakoterapii zaburzeń zachowania i snu u dzieci. *Forum Medycyny Rodzinnej*. 2011;5(5):424–428. <https://journals.viamedica.pl/forum-medycyny-rodzinnej/article/download/14160>. Accessed on November 6, 2019.
32. Kohanim S, Palioura S, Saeed HN, et al. Stevens–Johnson syndrome/toxic epidermal necrolysis: A comprehensive review and guide to therapy. I. Systemic disease. *Ocul Surf*. 2016;14(1):2–19. doi:10.1016/j.jtos.2015.10.002
33. Del Pazzo-Magana BR, Lazo-Langer A, Carleton B, Castro-Pastrana LL, Rieder MJ. A systematic review of treatment of drug-induced Stevens–Johnson syndrome and toxic epidermal necrolysis in children. *J Popul Ther Clin Pharmacol*. 2011;18:e121–133.
34. Ferrandiz-Pulido C, Garcia-Patos V. A review of causes of Stevens–Johnson syndrome and toxic epidermal necrolysis in children. *Arch Dis Child*. 2013;98(12):998–1003. doi:10.1136/archdischild-2013-303718
35. Abela C, Hartmann CE, De Leo A, et al. Toxic epidermal necrolysis (TEN): The Chelsea and Westminster Hospital wound management algorithm. *J Plast Reconstr Aesthet Surg*. 2014;67(8):1026–1032. doi:10.1016/j.bjps.2014.04.003
36. Mayes T, Gottschlich M, Khoury J, Warner P, Kagan R. Energy requirements of pediatric patients with Stevens–Johnson syndrome and toxic epidermal necrolysis. *Nutr Clin Pract*. 2008;23(5):547–550. doi:10.1177/0884533608323434
37. World Health Organization. WHO guidelines on the pharmacological treatment of persisting pain in children with medical illnesses. Geneva, Switzerland: World Health Organization: 2012. <https://apps.who.int/iris/handle/10665/44540>. Accessed on November 6, 2019.
38. Chave TA, Mortimer NJ, Sladden MJ, Hall AP, Hutchinson PE. Toxic epidermal necrolysis: Current evidence, practical management and future directions. *Br J Dermatol*. 2005;153(2):241–253. doi:10.1111/j.1365-2133.2005.06721.x
39. Olczak-Kowalczyk D, Krasuska-Sławińska E, Wieteska-Klimczak A, Siemieńska M, Daszkiewicz M, Ruszkowska L. Błona śluzowa jamy ustnej u dzieci z zespołem toksycznej nekrolizy naskórka. *Nowa Stomatologia*. 2011;1:23–30. http://www.nstomatologia.pl/wp-content/uploads/2014/10/ns_2011_023-030.pdf. Accessed on November 6, 2019.
40. McNichol L, Lund C, Ted Rosen T, Gray M. Medical adhesives and patient safety: State of the science consensus statements for the assessment, prevention, and treatment of adhesive-related skin injuries. *J Wound Ostomy Continence Nurs*. 2013;40(4):365–380. doi:10.1097/NOR.0b013e3182a39caf
41. Das S, Ramamoorthy R. Stevens–Johnson syndrome and toxic epidermal necrolysis in children. *Indian J Pediatr Dermatol*. 2018;19(1):9–14. doi:10.4103/ijpd.IJPD_120_17
42. Singer AJ, Talan DA. Management of skin abscesses in the era of methicillin-resistant *Staphylococcus aureus*. *N Engl J Med*. 2014;370(11):1039–1047. doi:10.1056/NEJMra1212788
43. Lin YS, Chen J, Li Q, Pan KP. Moisture vapour transmission rates of various transparent dressings at different temperatures and humidities. *Chin Med J (Engl)*. 2009;122(8):927–930. doi:10.1097/NOR.0b013e3182a39caf
44. Peng YZ, Yuan ZQ, Xiao GX. Effects of early enteral feeding on the prevention of enterogenic infection in severely burned patients. *Burns*. 2001;27(2):145–149. doi:10.1016/s0305-4179(00)00078-4