

Spina Bifida Guideline

Integument: Guidelines for the care of people with spina bifida

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Abstract.

PURPOSE: Skin-related issues have a significant impact on health, activities of daily living, and quality of life among people with spina bifida. Data presented by select clinics that participate in the National Spina Bifida Patient Registry reported that 26% of individuals had a history of pressure injuries with 19% having had one in the past year. The spina bifida community lack direct guidelines on prevention of these and other skin related issues. The Integument (skin) Guidelines focus on prevention, not treatment, of existing problems.

METHODS: Using a consensus building methodology, the guidelines were written by experts in spina bifida and wound care.

RESULTS: The guidelines include age-grouped, evidence-based guidelines written in the context of an understanding of the whole person. They are presented in table format according to the age of the person with spina bifida.

CONCLUSION: These guidelines present a standardized approach to prevention of skin-related issues in spina bifida. Discovering what results in successful minimization of skin-related issues with testing of technology or prevention strategies is the next step in protecting this vulnerable population.

Keywords: Spina bifida, skin integrity, pressure injury, pressure ulcer, prevention

1. Introduction

For people with spina bifida, skin-related issues have a significant impact on health, activities of daily living, and quality of life [1,2].

Data presented by select clinics that participate in the National Spina Bifida Patient Registry (NSBPR) indicate that 26% of individuals had a history of pressure injuries and 19% reported having had one in the past year [3]. Complications related to wounds were reported as the second most common primary diagnosis in spina bifida clinics [4]. The literature on this topic indicates that the cost to care for an individual

patient with a pressure injury ranges from \$20,900 to \$151,700 per pressure injury [5]. A multi-clinic study from the NSBPR identified seven factors associated with pressure injuries that included the level of lesion, wheelchair use, urinary incontinence, shunt presence, above the knee orthopedic surgery, recent surgery and male gender [3].

When comparing spina bifida patient admissions over a five-year period, with or without pressure injury, it was found that the average cost of hospitalization increased by 10% [6]. At the same time, the estimated average length of stay increased by 24% in the presence of pressure injuries among hospitalized patients with spina bifida, compared with their peers without these injuries [6]. These hospitalization statistics further emphasize the importance of attention to skin integrity in a coordinated care plan within the spina bifida population.

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Table 1
Clinical questions that informed the integument guidelines

Age group (from guidelines)	Clinical questions
0–11 months	– What is the evidence for the pathogenesis of skin breakdown (pressure injury) in infants with spina bifida? – Is there evidence that insensate skin in infants with spina bifida can be protected from breakdown?
1–2 years 11 months	– What is the evidence that early intervention and education will reduce skin injury?
3–5 years 11 months	– What activities promote self-awareness and self-inspection in children with spina bifida?
6–12 years 11 months	– What evidence is there that coaching independence will reduce skin breakdown?
13–17 years 11 months	– What are the key factors associated with skin breakdown?
and 18+ years	– Does the incidence of skin breakdown relate to the level of spina bifida? – What evidence is there that specific prevention measures will reduce the chance of skin breakdown?

Although skin issues are not confined to pressure injuries, pressure injury prevention programs have shown as much as a 67% reduction in incidence with a substantial reduction in the cost of care [1]. With that goal in mind, the information campaign to improve skin care awareness and wound prevention, “Did You Look?” is being evaluated as a prevention program [7]. Elements from this campaign are included in these guidelines.

These Integument (skin) Guidelines focus on prevention of skin issues and not treatment of existing problems. Though spina bifida specific evidence is limited, practices related to wound prevention in other populations can be applied to the spina bifida population. These guidelines are built on that evidence as well as clinical experiences of health care professionals in the spina bifida community.

1.1. Guidelines goals and outcomes

The desired clinical outcomes are clinically based on achieving the best skin health with consideration of the function of the skin along with quality of life that can be affected by loss of skin integrity. The goals of the Integument Guidelines are both practical and aspirational. Practical in the delineation of specific steps to minimize disruptions in skin integrity, and aspirational in our goal to dramatically reduce the incidence of pressure injury and thus have a significantly positive outcome on the health and financial impact on individuals with spina bifida.

Outcomes of the guidelines are written as levels of prevention. For the Integument Guidelines, the following three outcomes were identified.

Primary (intervene before an adverse health effect occurs)

- Maximize healthy skin, minimize disruptions in skin integrity.

Secondary (screen and identify problems in the earliest stages)

- Increase awareness of skin issues, risks, self-assessment, and prevention measures.

Tertiary (attempt to improve quality of life and reduce the symptoms)

- Improve health outcomes with minimal skin integrity issues across the lifespan.

2. Methods

The methodology for developing these guidelines has been published by Dicianno et al. [8]. The process included one hundred spina bifida experts from around the world who used a consensus building methodology.

A set of clinical questions was the foundation of the Integument Guidelines. These clinical questions were used to focus attention on the outcomes or goals of the guidelines. Table 1 shows clinical questions specific to five sequential age groups. It was these questions that informed the research and writing of the guidelines.

3. Results

The guidelines are presented in Table 2. The first column are the guidelines for skin care. The literature supporting the guidelines is listed in the column on the right. Each age group is represented with consideration to the potential variations in each individual’s functional abilities. New research which supports the guidelines has been added to this table. Note that some of the guidelines are cross-referenced to other guideline chapters.

4. Discussion

The guidelines for integument care provide a rational and comprehensive approach to the prevention of skin

Table 2
Integument (skin) guidelines with relevant citations as evidence

Age group	Guidelines	Evidence
0–11 months	1. Discuss insensate skin with parents and caregivers.	[9]
	2. Discuss the risk factors that may contribute to impaired skin integrity.	[3, 7, 9–15]
	3. Teach parents and caregivers to inspect the skin (especially weight bearing or insensate areas) for changes in color, texture, and temperature.	[4, 7, 9–15]
	4. Discuss the need to check water temperature and encourage the use of a bath water thermometer.	[7, 9–12]
	5. Suggest that parents and caregivers check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.	[7, 9–11]
	6. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.	[7, 11, 12, 16]
1–2 years, 11 months	1. Teach parents and caregivers to inspect the skin (especially weight bearing or insensate areas) for changes in color, texture, and temperature.	[3, 4, 7, 9–16]
	2. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.	[7, 11, 12, 16]
	3. Discuss the need to check water temperature and encourage the use of a bath water thermometer.	[7, 9–12]
	4. Suggest that parents and caregivers check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.	[7, 15, 16]
	5. Teach parents and caregivers how to inspect for well-fitting orthoses.	[3, 7, 16]
	6. Teach parents and caregivers that the child should wear protective clothing and footwear over insensate areas.	[7, 15, 16]
3–5 years, 11 months	7. Suggest that parents and caregivers seek treatment if the child's skin is compromised.	[7, 16]
	1. Teach parents and caregivers to inspect the skin daily (especially weight bearing or insensate areas) for changes in color, texture, and temperature.	[3, 4, 7, 9–16]
	2. Encourage the child's involvement in skin inspection.	[7, 10–16]
	3. Teach child to develop awareness of insensate areas.	[7, 10–16]
	4. Review with parents and caregivers the consequences of heat, moisture, or pressure to insensate areas.	[7, 10–16]
	5. Recommend the use of barrier creams to protect the skin from damage as a result of bowel and bladder incontinence.	[7, 10–16]
	6. Discuss the need to check water temperature and encourage the use of a bath water thermometer.	[7, 10–17]
	7. Suggest that parents and caregivers check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.	[7, 10–17]
	8. Teach parents and caregivers how to inspect for well-fitting orthoses.	[7, 9–12]
	9. Teach parents and caregivers that the child should wear protective clothing and footwear (including water shoes in a pool or on pool deck) over insensate areas.	[7, 15, 16]
6–12 years, 11 months	10. Suggest that parents and caregivers seek treatment if the child's skin is compromised.	[7, 16]
	1. Teach parents and caregivers to inspect the skin daily (especially weight bearing or insensate areas) for changes in color, texture, and temperature.	[3, 4, 7, 9–16]
	2. Encourage the child's involvement in skin inspection.	[7, 9–16]
	3. Teach child to develop awareness of insensate areas.	[7, 10–16]
	4. Review with parents and caregivers the consequences of heat, moisture, or pressure to insensate areas.	[7, 10–16]
	5. Teach parents and caregivers how to look for well-fitting orthoses and other equipment that may cause injury to skin.	[7, 15, 16]
	6. Teach parents and caregivers that the child should wear protective clothing and footwear over insensate areas.	[7, 10, 16]
	7. Discuss the need to check water temperature and encourage the use of a bath water thermometer.	[7, 10–16]
	8. Suggest that parents and caregivers check for hot surfaces that have been exposed to the sun such as car seats and playground equipment.	[7, 10–16]
	9. Promote adequate hydration and proper nutrition for healthy skin.	[7, 14, 18, 19], Nutrition, Metabolic Syndrome, and Obesity Guidelines
	10. Encourage parents, caregivers, and the child to keep skin clean and dry.	[7, 9–14]
	11. Suggest wearing seamless socks that are clean and dry.	[7, 12, 14]
	12. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.	[7, 16]
	13. Encourage parents and caregivers seek treatment if the child's skin is compromised.	[7, 12]
14. Advise parents and caregivers to engage non-ambulatory children in pressure relieving activities every 15 minutes.	[7, 10–11, 16–17]	

Table 2, continued

Age group	Guidelines	Evidence
13–17 years, 11 months and 18+ years	1. Inspect skin daily. Explore the teen perceptions of self-efficacy for skin checks and barriers to skin checks. Develop plans to increase self-efficacy, if needed.	[3, 4, 7, 10–16]
	2. Suggest children and adults who use wheelchairs to use a pressure-relieving cushion and check it daily.	[7, 12–16, 19]
	3. Identify and discuss risk factors that specifically increase the risk of pressure injuries in children and adults with spina bifida, such as using a wheelchair, having had surgery above the knee, shunts, a higher level of lesion, recent surgery, bladder incontinence, and being of the male gender.	[3, 7, 9–12, 14–16]
	4. Review with the caregiver, child, or adult the consequences of heat, moisture, or pressure to insensate areas.	[7, 9–17]
	5. Teach parents, caregivers, child, adult how to inspect for well-fitting orthoses.	[7, 9–17]
	6. Discuss the need to check water temperature and encourage the use of a bath water thermometer.	[7, 9–17]
	7. Suggest that children and adults check for hot surfaces that have been exposed to the sun such as car seats.	[7, 10–16]
	8. Promote adequate hydration and proper nutrition for healthy skin.	[7, 14, 18–19], Nutrition, Metabolic Syndrome, and Obesity Guidelines
	9. Encourage parents, caregivers, children, and adults to keep skin clean and dry.	[7, 9–14]
	10. Suggest wearing seamless socks that are clean and dry.	[7, 12, 14]
	11. Suggest the use of antiperspirant on areas with perspiration, such as the feet and intertriginous areas.	[7, 16]
	12. Seek treatment if the skin is compromised.	[7, 12]
	13. Advise children and adults who are non-ambulatory and use a wheelchair to engage in pressure-relieving activities every 15 minutes.	[7, 10–11, 16–17]
	14. Teach safe transfer skills to non-ambulatory patients.	[7, 14]
	15. Seek treatment immediately for any pressure injury. Refer to wound clinic for any pressure injury at stage three or greater.	[1, 5, 7, 12]

integrity issues in the spina bifida population. Given the large number of people with spina bifida who present with a pressure injury, prevention is a priority [20]. These guidelines provide a rational and effective approach to preventing or minimizing integument issues.

We know from previous work what factors are associated with an increased risk of pressure injury [3]. So what more can be done? There has been interval work since the release of the guidelines that has strived to improve outcomes in skin integrity. Studies focusing on evidence-based wellness programs, mobile health, applications for smart phone applications, and other novel technologies have shown improved health regarding preventable secondary conditions in spina bifida [21] as well as short term positive changes in self-management and independence [22]. In one such system, a smart-phone application allowed the uploading photos of pressure injuries through a secure system [22]. Studies utilizing common technology can greatly enhance evaluation, diagnosis, and initiation of care.

This is further reinforced by a review article looking at the potentially positive effect of compensatory feedback technologies [23]. Although this article addressed spinal cord injury patients, there is considerable overlap in the literature on wheelchair users between spina bifida and spinal cord injury. With varied feedback technologies included, the potential posi-

tive effect of compensatory feedback and technology to guide behaviors that reduce the risk of pressure injury is significant [23].

The wound care literature supports a standardized approach that is important for pressure injury prevention which includes risk assessment, evidence-based guidelines, prevention strategies, and care specific to vulnerable populations. The “Did You Look” bundle of risk assessment, skin assessment, and education is one such approach and is currently being tested through select National Spina Bifida Patient Registry clinics [7].

In summary, these guidelines provide levels of prevention strategies customized to specific age groups that promote skin integrity across the lifespan of individuals with spina bifida.

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The Spina Bifida Association has already embarked on a systematic process for reviewing and updating the guidelines. Future guidelines updates will be made available as they are completed.

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Conflict of interest

There are no conflicts represented by the authors.

References

- [1] Ottolini K, Harris AB, Amling JK, Kennelly AM, Phillips LA, Tosi LL. Wound care challenges in children and adults with spina bifida: an open-cohort study. *J Pediatr Rehabil Med*. 2013; 6(1): 1–10. doi: 10.3233/PRM-130231.
- [2] Clark M. Pressure ulcers and quality of life. *Nursing Standard* (through 2013). 2002 Feb 13; 16(22): 74.
- [3] Kim S, Ward E, Dicianno BE, Clayton GH, Sawin KJ, Beierwaltes P, et al. Factors associated with pressure ulcers in individuals with spina bifida. *Arch Phys Med Rehabil*. 2015 Aug; 96(8): 1435–1441.e1. doi: 10.1016/j.apmr.2015.02.029.
- [4] Curley MAQ, Razmus IS, Roberts KE, Wypij D. Predicting pressure ulcer risk in pediatric patients: the Braden Q Scale. *Nurs Res*. Jan-Feb 2003; 52(1): 22–33. doi: 10.1097/00006199-200301000-00004.
- [5] Agency for Healthcare Research and Quality. Rockville, MD. Preventing Pressure Ulcers in Hospitals. 2014 Oct. Available from <https://www.ahrq.gov/patient-safety/settings/hospital/resource/pressureulcer/tool/index.html>.
- [6] Wang Y, Ouyang L, Dicianno BE, Beierwaltes P, Valdez R, Thibadeau J, et al. Differences in length of stay and costs between comparable hospitalizations of patients with spina bifida with or without pressure injuries. *Arch Phys Med Rehabil*. 2019 Aug; 100(8): 1475–1481. doi: 10.1016/j.apmr.2018.12.033.
- [7] Spina Bifida Association (Internet). Did You Look? Skin integrity bundle; 2017. Available from <https://www.spinabifidaassociation.org/about-our-research/did-you-look/>.
- [8] Dicianno BE, Beierwaltes P, Dosa N, Raman L, Chelliah J, Struwe S, et al. Scientific methodology of the development of the guidelines for the care of people with spina bifida: an initiative of the spina bifida association. *Disabil Health J*. 2020 Apr; 13(2): 100816. doi: 10.1016/j.dhjo.2019.06.005.
- [9] Baharestani MM, Ratliff CR. Pressure ulcers in neonates and children: an NPUAP white paper. *Adv Skin Wound Care*. 2007 Apr; 20(4): 208, 210, 212, 214, 216, 218–20. doi: 10.1097/01.ASW.0000266646.43159.99.
- [10] Mahmood D, Dicianno B, Bellin M. Self-management, preventable conditions and assessment of care among young adults with myelomeningocele. *Child Care Health Dev*. 2011 Nov; 37(6): 861–5. doi: 10.1111/j.1365-2214.2011.01299.x.
- [11] Schlüer AB. Pressure ulcers in maturing skin – a clinical perspective. *Journal of Tissue Viability*. 2017; 26(1): 2–5. doi: 10.1016/j.jtv.2016.10.001.
- [12] Black JM, Edsberg LE, Baharestani MM, Langemo D, Goldberg M, McNichol L, et al. Pressure ulcers: avoidable or unavoidable? Results of the national pressure ulcer advisory panel consensus conference. *Ostomy Wound Manage*. 2011 Feb 1; 57(2): 24–37.
- [13] Brustrom J, Thibadeau J, John L, Liesmann J, Rose S. Care coordination in the spina bifida clinic setting: current practice and future directions. *J Pediatr Health Care*. Jan-Feb 2012; 26(1): 16–26. doi: 10.1016/j.pedhc.2010.06.003.
- [14] DeJong G, Hsieh CHJ, Brown P, Smout RJ, Horn SD, Ballard P, et al. Factors associated with pressure ulcer risk in spinal cord injury rehabilitation. *Am J Phys Med Rehabil*. 2014 Nov; 93(11): 971–86. doi: 10.1097/PHM.0000000000000117.

- [15] Lala D, Dumont FS, Leblond J, Houghton PE, Noreau L. Impact of pressure ulcers on individuals living with a spinal cord injury. *Arch Phys Med Rehabil*. 2014 Dec; 95(12): 2312–9. doi: 10.1016/j.apmr.2014.08.003.
- [16] Kottner J, Cuddigan J, Carville K, Balzer K, Berlowitz D, Law S, et al. Prevention and treatment of pressure ulcers/injuries: the protocol for the second update of the international Clinical Practice Guideline 2019. *J Tissue Viability*. 2019 May; 28(2): 51–58. doi: 10.1016/j.jtv.2019.01.001.
- [17] Bryant RA, Nix DP. *Acute and chronic wounds: Current management concepts*. 5th ed. St. Louis: Elsevier; 2016. Chapter 8, Developing and maintaining a pressure ulcer prevention program, 140–143.
- [18] Dicianno BE, Wilson R. Hospitalizations of adults with spina bifida and congenital spinal cord anomalies. *Arch Phys Med Rehabil*. 2010 Apr; 91(4): 529–35. doi: 10.1016/j.apmr.2009.11.023.
- [19] Dorner B, Posthauer ME, Thomas D. The role of nutrition in pressure ulcer prevention and treatment: National Pressure Ulcer Advisory Panel white paper. *Adv Skin Wound Care*. 2009 May; 22(5): 212–21. doi: 10.1097/01.ASW.0000350838.11854.0a.
- [20] Delmore B, Deppisch M, Sylvia C, Luna-Anderson C, Nie AM. Pressure injuries in the pediatric population: a national pressure ulcer advisory panel white paper. *Adv Skin Wound Care*. 2019 Sep; 32(9): 394–408. doi: 10.1097/01.ASW.0000577124.58253.66.
- [21] Dicianno BE, Lovelace J, Peele P, Fassinger C, Houck P, Bur-sic A, et al. Effectiveness of a wellness program for individuals with spina bifida and spinal cord injury within an integrated delivery system. *Arch Phys Med Rehabil*. 2016 Nov; 97(11): 1969–1978. doi: 10.1016/j.apmr.2016.05.014.
- [22] Dicianno BE, Fairman AD, McCue M, Parmanto B, Yih E, Mc-Coy A, et al. Feasibility of using mobile health to promote self-management in spina bifida. *Am J Phys Med Rehabil*. 2016 Jun; 95(6): 425–37. doi: 10.1097/PHM.0000000000000400.
- [23] Vos-Draper TL, Morrow MMB. Seating-related pressure injury prevention in spinal cord injury: a review of compensatory technologies to improve in-seat movement behavior. *Curr Phys Med Rehabil Rep*. 2016 Dec; 4(4): 320–328. doi: 10.1007/s40141-016-0140-7.