Pediatric **Dermatology**

Content Strategist: Belinda Kuhn

Content Development Specialist: Sharon Nash

Project Manager: Sruthi Viswam

Design: Christian Bilbow

Illustration Manager: Jennifer Rose

Illustrator: Richard Tibbits Marketing Manager: Katie Alexo

Pediatric **Dermatology**

FOURTH EDITION

Bernard A. Cohen MD

Director of Pediatric Dermatology Johns Hopkins Children's Center Professor of Pediatrics and Dermatology Johns Hopkins University School of Medicine Baltimore Maryland USA

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Preface

Since I began taking clinical photographs during my residency training over 30 years ago, I have been impressed by the virtually unlimited variation in the expression of skin disease. However, with careful observation, clinical patterns that permit the development of a reasonable differential diagnosis emerge. In the fourth edition, I have been able to use over 600 images, a third of which are new, to demonstrate the diverse variations and common patterns that are fundamental to an understanding of skin eruptions in children. Moreover, since I have catalogued all new images on our online dermatology webAtlas at dermatlas.org, which is the source of most of the new images in this edition, the reader is referred to this site to view additional images and more detailed discussion of specific clinical cases. There are over 500 contributors to dermAtlas, and you are invited to participate.

Pediatric Dermatology is designed for the pediatric and primary care provider with an interest in dermatology and the dermatology practitioner who cares for children. The text is organized around practical clinical problems, and most chapters end with an algorithm for developing a differential diagnosis. This book should not be considered an encyclopedic text of pediatric dermatology; it should be used in conjunction with the further reading suggested at the end of Chapter 1. Classic papers and more recent literature are included in the further reading lists at the end of each chapter.

At Hopkins, we have been fortunate to have oral pathologists on the dermatology faculty in the roles of teacher and consultant. With their help, the importance of recognizing oral lesions in the care of children is reflected in Chapter 9, which is devoted to oral pathology. Although the focus of this chapter is on primary lesions of the oral mucosa, a discussion of clues of systemic disease is included. Chapter 2, which is devoted to dermatologic disorders of newborns and infants, remains the longest chapter in the book due to the continued blossoming of neonatology as a respected pediatric discipline. I never cease to be amazed by how human beings manipulate their skin accidentally, deliberately, secretly, and/or therapeutically. With this in mind, Chapter 10, Factitial Dermatoses, concludes with several disorders that are triggered, exacerbated, or caused primarily by external factors.

Finally, the format of the text should be user-friendly. The pages and legends have been numbered in a standard textbook fashion, and the index is again revised to include all of the disorders listed in the text as well as the legends. The text and images incorporate advances made in diagnosis, evaluation, and treatment during the last 7 years, since the publication of the third edition. I only hope that students of pediatric dermatology will enjoy reading the book as much as I enjoyed writing and illustrating it.

Bernard A. Cohen 2012

Acknowledgments

This book would not have been possible without the help of the children and parents who allowed me to photograph their skin eruptions, and the practitioners who referred them to me. I am particularly indebted to the faculty, residents, nurse practitioners, nurses, physicians assistants, and students at the Johns Hopkins Children's Center and the Departments of Pediatrics and Dermatology at the Johns Hopkins University School of Medicine for their inspiration and support. I would again like to thank my friends at the Children's Hospital of Pittsburgh where this book was first conceived.

I have a new group of over 500 friends and contributors, most of whom I have met through an ever-expanding online dermatology image project 'dermatlas'. My association with dermatlas as one of the founding Editors gives me access to an incredible national and international repository of cutaneous images. It was my honor to work with Editor emeritus and medical informatics maven Christoph Lehmann, a neonatologist who probably knows more dermatology than any other neonatologist in the country. My son, Michael Cohen, a young and rising computer science wizard is in the process of rewriting and modernizing the platform, which hopefully will be completed by the time this new edition of Pediatric Dermatology is published.

I am also indebted to the oral pathology faculty at Hopkins who call dermatology their home. They have taught me to seek clues for dermatologic and systemic disease from evaluation of the mucous membranes, and to respect oral pathology in its own right. Without them, the conception of Chapter 9 and the most recent updates would not have been possible.

I continue to be grateful for the persistent prodding and sensitive guidance of the editors at Elsevier who are responsible for completion of this book in a timely fashion. I would also like to thank Tracy Shuford for keeping the lines of communication open between the Publisher and my office, despite the 6-hour time difference.

Special thanks go to Kate Puttgen, my colleague in crime in pediatric dermatology at the Children's Center; John Mavrolopoulos, a rising dermatology resident star at the School of Medicine, and my wife Sherry Cohen, Family Practice Nurse Practitioner, who still works in dermatology in spite of me, and all of whom have contributed significantly to this edition.

I would like to thank the residents in dermatology and pediatrics, who by their questions and consultations, have helped me prioritize topics for inclusion in this book.

Finally, I would like to again acknowledge Dr Nancy Esterly, who contributed the foreword to the second edition (reprinted in the third edition). I think of her often and would like to honor her by using her foreword in this edition as well. Dr Esterly taught me that pediatric dermatology could be exciting and academically challenging. As a role model and friend, she continues to guide all of us in pediatric dermatology. I would also like to acknowledge Dr Frank Oski who brought me home to Baltimore, where he incorporated pediatric dermatology into the pediatric training program. Hopefully, we can live up to the high standards which he demanded.

Figure Credits

The following figures have been reprinted from Zitelli BJ, Davis HW (eds). Atlas of pediatric physical diagnosis, 3rd edn. Mosby, St Louis, 1997:

4.10, 7.8, 7.9, 8.1, 8.15, 8.49, 10.5, 10.7, 10.8, 10.11, 10.13

I am grateful for the use of images from: www.dermatlas.org, and to Dr Russ Corio and Dr Gary Warnock Associate Professor of Dermatology at the Johns Hopkins University School of Medicine, for contributing additional images to the chapter on the Oral Cavity (Ch. 9).

Dedication

To Sherry for her continued patience, love, understanding, and encouragement during the revision of this book, which took longer than I thought!

To Michael, Jared, and Jennie for keeping me young and laughing. It has been exciting to see them mature into young adults who now contribute to the care of children and adults in their own ways.

To all of the children who made this project possible.

Foreword

NOTE FROM DR COHEN

I have asked the managing editor to reprint the Foreword from the second edition (also reprinted in the third edition) written by Dr. Nancy Esterly to honor her for her contributions to pediatric dermatology, the training of many practitioners of the specialty, and my own career. In the spring of 1983 when I was desperately searching for a mentor in pediatric dermatology, Nan adopted me during my elective month at Childrens's Memorial Hospital in Chicago.

Dr. Esterly has been the quintessential practitioner of pediatric dermatology since her pediatric and dermatology training in Baltimore over 40 years ago. She was one of the founders of the Society for Pediatric Dermatology and embodies the tripartite mission of pediatric dermatology of patient care, resident teaching, and clinical research.

FOREWORD TO THE SECOND EDITION

It isn't often that one encounters a single author textbook that is outstanding in both text and illustrations. But, once again, Bernard Cohen has crafted an exceptional basic pediatric dermatology text liberally illustrated with photographs depicting a wide range of skin problems in infants and children.

In this fourth edition of Pediatric Dermatology, the text has been expanded to include a 20 page chapter devoted entirely to mucosal lesions and accompanied by more than 50 new photographs of patients with problems ranging from the common herpes simplex infection to the uncommon ectodermal dysplasias. In keeping with the very successful style of previous editions, the requisite algorithm, diagrams of the oral cavity and up-to-date references are included in this chapter. In addition, new photographs have been added and some old ones replaced throughout the book.

For beginners in this discipline, Dr. Cohen's text is an excellent place to start. For those of us who practice pediatric dermatology, there is still much to be learned from a well-put-together text such as this one.

Nancy B. Esterly, M.D.
Professor Emeritus
Medical College of Wisconsin
Milwaukee, Wisconsin

Contributors

Anna M. Bender MD

Assistant Professor of Dermatology Department of Dermatology Weill Cornell Medical College New York, NY, USA

Sherry Guralnick Cohen CRNP-F APRN-PMH

Family Nurse Practitioner, Dermatology Pikesville, MD, USA

John C. Mavropoulos MD MPH PhD

Resident in Dermatology
Department of Dermatology
Johns Hopkins University School of Medicine
Baltimore, MD, USA

Katherine B. Püttgen MD

Assistant Professor Department of Dermatology Johns Hopkins University School of Medicine Baltimore, MD, USA

Introduction to Pediatric Dermatology

Bernard A. Cohen

ANATOMY OF THE SKIN

Most of us think of skin as a simple, durable covering for the skeleton and internal organs. Yet skin is actually a very complex and dynamic organ consisting of many parts and appendages (Fig. 1.1). The outermost layer of the epidermis, the stratum corneum, is an effective barrier to the penetration of irritants, toxins, and organisms, as well as a membrane that holds in body fluids. The remainder of the epidermis, the stratum granulosum, stratum spinosum, and stratum basale, manufactures this protective layer. Melanocytes within the epidermis are important for protection against the harmful effects of ultraviolet light, and the Langerhans cells and other dendritic cells are one of the body's first lines of immunologic defense and play a key role in systemic and cutaneous diseases such as drug reactions.

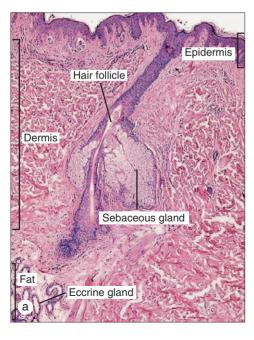
The dermis, consisting largely of fibroblasts and collagen, is a tough, leathery, mechanical barrier against cuts, bites, and bruises. Its collagenous matrix also provides structural support for a number of cutaneous appendages. Hair, which grows from follicles deep within the dermis, is important for cosmesis, as well as protection from sunlight and particulate matter. Sebaceous glands arise

as an outgrowth of the hair follicles. Oil produced by these glands helps to lubricate the skin and contributes to the protective function of the epidermal barrier. The nails are specialized organs of manipulation that also protect sensitive digits. Thermoregulation of the skin is accomplished by eccrine sweat glands as well as changes in the cutaneous blood flow regulated by glomus cells. The skin also contains specialized receptors for heat, pain, touch, and pressure. Sensory input from these structures helps to protect the skin surface against environmental trauma. Beneath the dermis, in the subcutaneous tissue, fat is stored as a source of energy and also acts as a soft protective cushion.

EXAMINATION AND ASSESSMENT OF THE SKIN

The skin is the largest, and most accessible and easily examined organ of the body, and it is the organ of most frequent concern to the patient. Therefore, all practitioners should be able to recognize basic skin diseases and dermatologic clues to systemic disease.

Optimal examination of the skin is best achieved in a well-lit room. The clinician should inspect the entire skin surface, including the hair, nails, scalp, and mucous membranes. This may present



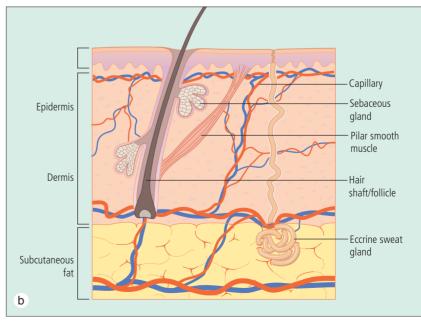


Fig 1.1 (a) Skin photomicrograph and (b) schematic diagram of normal skin anatomy.



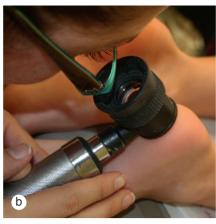




Fig 1.2 Examination of pigmented nevi with (a) DermLite II Hybrid by 3Gen, (b) Welch Allyn Episcope Skin Surface Microscope, and (c) Welch Allyn Otoscope.

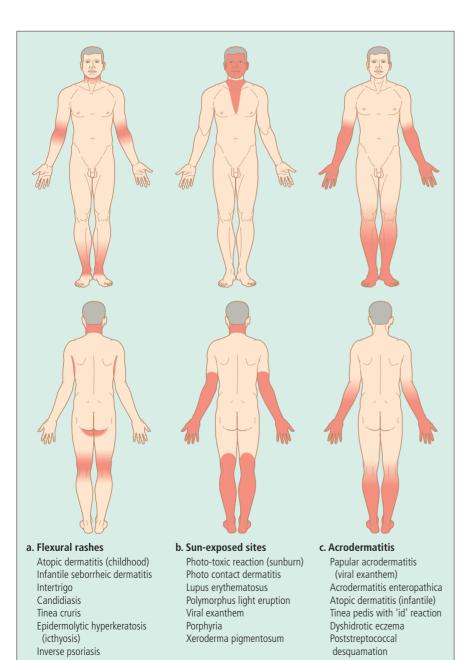


Fig 1.3 (a-h) Pattern diagnosis.

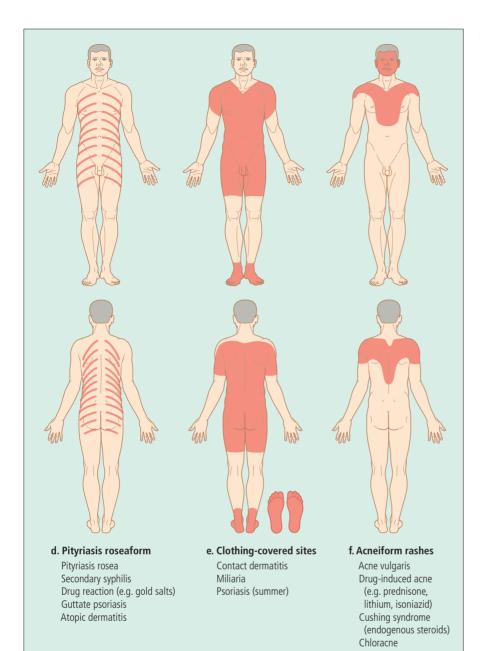


Fig 1.3 Continued.

particular problems in infants and teenagers, since it may be necessary to examine the skin in small segments to prevent cooling or embarrassment, respectively. Although no special equipment is required, a hand lens and side-lighting are useful aids in the assessment of skin texture and small discrete lesions. In many offices, the otoscope can be adapted for this purpose by removing the plastic speculum.

There are also a number of relatively inexpensive portable dermatoscopic devices, which can also be used to enhance the examination (also known as epiluminescence microscopy). These instruments have traditionally provided a magnified (×10) view of the skin with a non-polarized light source, a transparent plate, and a liquid medium between the dermatoscope and the skin. This allows for a view of the skin without interference from surface reflections. Dermatoscopic heads can be purchased for otoscope/ ophthalmoscope handpieces, and mineral oil or alcohol gel can be applied directly to the skin lesion. Newer devices achieve the same view without the liquid medium by using polarized light (Fig. 1.2).

Despite the myriad of conditions affecting skin, a systematic approach to the evaluation of a rash facilitates and simplifies the process of developing a manageable differential diagnosis. After assessing the general health of a child, the practitioner should obtain a detailed history of the cutaneous symptoms, including the date of onset, inciting factors, the evolution of lesions, and the presence or absence of pruritus. Recent immunizations, infections, drugs, and allergies may be directly related to new rashes. The family history may suggest a hereditary or contagious process, and the clinician may need to examine other members of the family. A review of nursery records and photographs will help to document the presence of congenital lesions.

Attention should then turn to the distribution and pattern of the rash. The distribution refers to the location of the skin findings, while the pattern defines a specific anatomic or physiologic arrangement. For example, the distribution of a rash may include the extremities, face, or trunk, while the pattern could be flexural or intertriginous areas (Fig. 1.3a). Other common patterns include

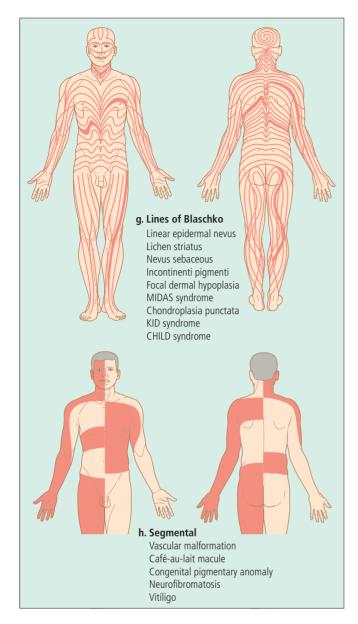


Fig 1.3 Continued.

sun-exposed sites, acrodermatitis (predilection for the distal extremities), pityriasis rosea (truncal, following the skin cleavage lines), clothing-protected sites, acneiform rashes, Blaschko lines, and segmental lesions (Fig. 1.3b-g).

Next, the clinician should consider the local organization and configuration of the lesions, defining the relationship of primary and secondary lesions to one another in a given location (Table 1.1) and the shape of the lesions. Are the lesions diffusely scattered or clustered (herpetiform)? Are they dermatomal, linear, serpiginous, circular, annular, or reticulated?

The depth of the lesions in the skin, as noted by both observation and palpation, may also give further clues (Table 1.2). Disruption of the normal skin markings by scale, papules, vesicles, or pustules points to the involvement of the epidermis. Alterations in skin color alone can occur in epidermal and dermal processes. In disorders of pigmentation, the color of the pigment may suggest the anatomic depth of the lesion. Shades of brown are present in flat junctional nevi, lentigines, and café-au-lait spots, where the increased pigment resides in the epidermis or superficial dermis. In Mongolian spots

and nevus of Ota, the Tyndall effect results in bluish-green to gray macules from melanin in the mid-dermis. If the epidermal markings are normal but the lesion is elevated, the disorder usually involves the dermis. Dermal lesions have well-demarcated firm borders. Nodules and tumors deep in the dermis or subcutaneous tissue can distort the surface markings, which are otherwise intact. Some deep-seated lesions can only be appreciated by careful palpation.

Lesion color can provide important clues for diagnosis and the pathophysiology of the underlying process (Table 1.3). Brown, blue, gray, bronze and black lesions are associated with disorders that alter normal pigmentation, while white lesions may be associated with loss of normal pigmentation or the accumulation of scale, crust, or exudates. Red and blue lesions are associated with inflammatory and vascular processes. Non-blanching blue or purple lesions should suggest the presence of purpura. Yellow lesions occur when the skin is infiltrated with inflammatory or tumor cells containing lipid. Other pigments from topical agents (e.g. silver, gold), oral medications (e.g. minocycline, amiodarone), foreign bodies (e.g. asphalt, tattoo pigments), and infectious agents (e.g. Pseudomonas species, Corynebacterium species) may impart specific colors to cutaneous lesions.

Finally, the clinician may develop a differential diagnosis using the morphology of the cutaneous lesions. Primary lesions (macules, papules, plaques, vesicles, bullae, pustules, wheals, nodules, and tumors) arise de novo in the skin (Fig. 1.4). Secondary lesions (scale, crust, erosions, ulcers, scars with atrophy and/or fibrosis, excoriations, and fissures) evolve from primary lesions or result from scratching of primary lesions by the patient (Fig. 1.5).

The practitioner who becomes comfortable with dermatology will integrate all of these approaches into their evaluation of a child with a skin problem. This will be reflected in the clinically focused format of this text.

Each chapter will finish with an algorithm that summarizes the material in a differential diagnostic flow pattern. The limited bibliography includes comprehensive, historically significant, and/or well-organized reviews of the subject. Readers may also find some of the texts and online further reading listed at the end of this chapter useful.

DIAGNOSTIC TECHNIQUES

Potassium hydroxide preparation

There are a number of rapid, bedside diagnostic procedures in dermatology. One of the most useful techniques is a wet mount of skin scrapings for microscopic examination (Fig. 1.6). Potassium hydroxide (KOH), 20%, is used to change the optic properties of skin samples and make scales more transparent. The technique requires practice and patience.

The first step is to obtain the material by scraping loose scales at the margin of a lesion, nail parings, subungual debris, or the small pearly globules from a molluscum body. Short residual hair stubs (black dots in tinea capitis) may also be painlessly shaved off the scalp with a #15 blade. Scale is placed on the slide and moved to the center with a cover slip. One or two drops of KOH are added and gently warmed with a match or the microscope light. Boiling the specimen will introduce artifact and should be avoided. Excess KOH can be removed with a paper towel applied to the edge of the cover slip. Thick specimens may be more easily viewed after gentle, but firm pressure is applied to the cover slip with a pencil eraser. Thick scale will also dissolve after being set aside for 15-20 min.

View the preparation under a microscope, with the condenser and light at low levels to maximize contrast, and with the objective at ×10. Focus up and down as the entire slide is rapidly scanned.

Organization and configuration of lesions Filiform Linear **Serpiginous Annular** Herpetiform Reticulated (thread-like) Geographic **Dermatomal** Epidermal nevi Herpes zoster **Psoriasis** Ringworm Herpes simplex Cutis marmorata Wart Psoriasis Lichen striatus Vitiligo Erythema Granuloma infection Livedo reticularis Dermatosis Geographic Congenital Contact Nevus marginatum annulare Herpes zoster papulosa tongue dermatitis Cutaneous Subacute Dermatitis phlebectasia Nummular depigmentosus nigra Warts Reticulated and Becker nevus larvae cutaneous herpetiformis Syringocysteczema Ichthyosis Café-au-lait spot confluent Erythema migrans lupus adenoma **Psoriasis** Port-wine stain Elastosis Atopic papillomatosis papilliferum annulare Porokeratosis perforans dermatitis Erythema ab Skin tag centrifugum Pigmented Incontinentia serpiginosa Erythema igne annulare nevus pigmenti centrifugum Erythema chronicum migrans Erythema marginatum

Table 1.1 Organization and configuration of lesions

Anatomic depth of lesions			
Cutaneous structure	Physical findings	Specific skin disorder	
Epidermis	Altered surface markings Scale, vesicle, crust Color changes (black, brown, white)	Impetigo Café-au-lait spot Atopic dermatitis Vitiligo Freckle	
Epidermis + dermis	Altered surface markings Scale, vesicle, crust Distinct borders Color changes (black, brown, white, and/or red) Edema	Psoriasis Atopic dermatitis Cutaneous lupus erythematosus	
Dermis	Normal surface markings Color changes Altered dermal firmness	Urticaria Granuloma annulare Hemangioma Blue nevus	
Subcutaneous tissue	Normal surface markings Normal or red skin color Altered skin firmness	Hematoma Cold panniculitis Erythema nodosum	

Table 1.2 Anatomic depth of lesions

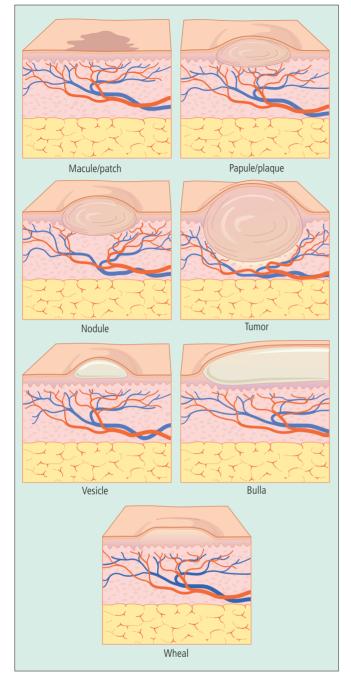


Fig 1.4 Primary skin lesions. Macule: a small (usually = 1 cm), flat lesion showing an alteration in color or tone. Large macule is a patch. Papule: a small (= 1 cm), sharply circumscribed, elevated lesion. An elevated lesion over 1 cm is referred to as a plaque. Nodule: a soft or solid mass in the dermis or subcutaneous fat. Tumor: a large nodule, localized and palpable, of varied size and consistency. Vesicle: a blister containing transparent fluid. Bulla: a large blister. Wheal: an evanescent, edematous, circumscribed elevated lesion that appears and disappears quickly. (Adapted from CIBA.)

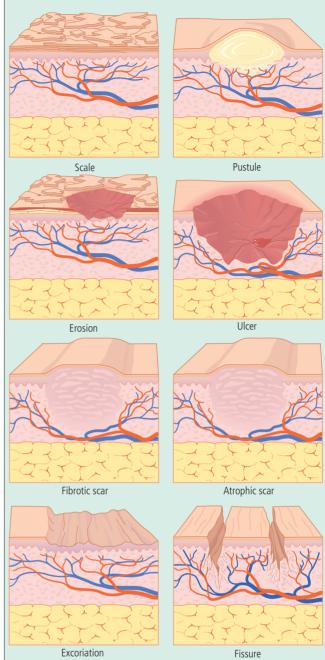


Fig 1.5 Secondary skin lesions. Scale: dry and/or greasy fragments of adherent epidermis. Pustule: a sharply circumscribed lesion containing free pus. Crust: a dry mass of exudate from erosions or ruptured vesicles/ pustules, consisting of serum, dried blood, scales, and pus. Erosion: welldefined partial-thickness loss of epidermis. Ulcer: a clearly defined, fullthickness loss of epidermis that may extend into the subcutis. Scar: a permanent skin change resulting from new formation of connective tissue after destruction of the epidermis and cutis. When the loss of dermis and/ or fat is prominent the lesion may be atrophic. Fibrosis may result in firm thickened papules or plaques. Excoriation: any scratch mark on the surface of the skin. Fissure: any linear crack in the skin, usually accompanied by inflammation and pain. (Adapted from CIBA.)