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3rd Edition

A Handbook of VITILIGO & Color Atlas





SK Punshi

Foreword
James J Nordlund

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CHAPTER

8

Classification

INTRODUCTION

The vitiligo patches are classified in different ways by different authors as follows:

- *Elmofty's classification (according to the extent of the involved area)*:
  - Vitiligo localization
  - Vitiligo generalist
  - Vitiligo universalis
- *Punshi's classification*:
  - Acute, subacute, and chronic vitiligo
  - Responsive to treatment and non-responsive or resistant
  - Special:
    - Recurrent vitiligo
    - Fleeting vitiligo
    - Linear vitiligo
    - Congenital vitiligo
- *Classification according to the distribution of lesion*:
  - Focal:
    - Localized
    - Segmental (dermatomal or zosteriform)
    - Mucosal
  - Generalized:
    - Acrofacial
    - Vitiligo
    - Universal
    - Trichrome vitiligo
    - Leukoderma acquisitum centrifugum (peri-halo nevus)
- *Etiological classification*:
  - Autoimmune vitiligo or progressive vitiligo
  - Segmental or dermatomal vitiligo
  - Chemical or contact vitiligo
- *Behl's classification*:
  - Active
  - Quiescent
  - Improving
  - Zosteriformis
- *Clinicopathological classification*:
  - Melanocytopenic leukoderma reduction or absence of melanocytes, e.g.:
    - Chemical leukoderma
    - Piebaldism, etc.
  - Melanopenic leukoderma—reduction or absence of melanin pigment, e.g.:
    - Albinism
    - Nevus depigmentosus
- *Etiopathological classification of vitiligo*:
  - Autoimmune vitiligo or progressive vitiligo, idiopathic vitiligo
  - Segmental or dermatomal vitiligo
  - Chemical or contact vitiligo
- According to the patterns of distribution of lesions, Dutta (1998) suggested the following three clinical types of vitiligo:
  i. Vitiligo vulgaris—53.5%
  ii. Vitiligo acro-orificialis—29.5%
  iii. Vitiligo segmentalis (zosteriforms)—70% dermatomal
- *Koga's classification*:
  - Focal vitiligo
  - Segmental vitiligo

Different pathomechanisms with yet unknown deep-seated implications are possibly involved. Vitiligo may be external manifestation linked with constitutional

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diathesis and/or systemic disorders determined fundamentally by genes.

- *Dutta AK feels inclined to suggest different subsets of vitiligo patients as*:
  - Neuropathic
  - Autoimmunopathic
  - Atopic
  - Idiopathic
- Such an approach in the total evaluation of a vitiligo patient will likely be of much help in the initial assessment of prognosis and in planning the therapeutic program.
- *Histological classification of vitiligo on the basis of dopa reaction*:
  - Absolute
  - Reactive type I
  - Relative type II

CLINICAL CLASSIFICATION OF VITILIGO: DISCUSSION AND CLARIFICATION

There have been many attempts to classify types of vitiligo, often with confusing results. One simple classification is presented here.

Localized

Focal

One or more macules in one area but not clearly in a segmental or zosteriform distribution.

Segmental

According to Koga, segmental vitiligo is characterized by the presence of one or more macules that involve only a unilateral segment of the body, such as part of the face, part of the trunk, one extremity, or part of an extremity. In this type of vitiligo, the lesions abruptly stop at the midline of the affected segment, creating a clear demarcation between the affected and unaffected areas.

Mucosal

Vitiligo of the mouth and mucous membranes, vitiligo on the genitalia.

Generalized

Acrofacial: Distal part of the extremities and face.

Vulgaris: Scattered macules.

Mixed: Acrofacial and vulgaris, or segmental and acrofacial and/or vulgaris.

Universal

Complete or nearly complete depigmentation characterizes the various types of vitiligo. The incidence of each type varies from study to study. Generalized vitiligo is reported to range from 50 to 90% in different studies. In a study by Song et al. (1994) with 1,315 patients, 50.2% of cases were classified as generalized vitiligo, comprising acrofacial (14.4%) and vulgaris (33.8%) subtypes. The remaining 49.7% were categorized as localized vitiligo, which included focal (33.7%), segmental (15.4%), and mucosal (0.6%) forms, with one case of universal type.

Localized vitiligo, except for the segmental type, may serve as an intermediary stage that could evolve into generalized vitiligo or, rarely, into universal vitiligo. However, segmental vitiligo is never an intermediary stage. There are very few patients who exhibit both generalized and segmental vitiligo. The authors are aware of two such individuals, and they seem to have two distinct types of vitiligo, indicating the coexistence of different patterns in rare cases.

Vitiligo can also be classified into three major clinical types, segmental, nonsegmental, and mixed, as follows:

- Segmental (unilateral)
- *Nonsegmental (bilateral)*:
  - Focal
  - Mucosal

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  - Acrofacial
  - Vulgaris
  - Universal
- Mixed, segmental, and nonsegmental

Segmental vitiligo is characterized by depigmented patches distributed on one side of the body in a pseudodermatomal pattern. In this type of vitiligo, the depigmented patches are confined to a specific area of the skin, and the central border of the vitiligo lesions does not cross the body's midline. This distinct distribution pattern sets segmental vitiligo apart from other types of vitiligo where lesions may be more generalized and involve both sides of the body.

CLASSIFICATION OF VITILIGO DEPENDING ON THE ACTIVITY OF DISEASE (BEHL) (TABLE 1)

Queer and Unpredictable Vitiligo Activity: Course of Vitiligo

The progression of vitiligo is often unpredictable. Typically, the disease follows a slow course, but it can also stabilize or worsen rapidly. The spread of vitiligo can occur through the appearance of new depigmented macules, the enlargement of existing lesions, or a combination of both. In rare instances, there is a rapid and extensive depigmentation over a few days to weeks, referred to as vitiligo fulminans. Conversely, spontaneous repigmentation can also occur, although it is less common. Spontaneous repigmentation is observed in approximately 10–20% of vitiligo patients, most frequently in sun-exposed areas, but it is usually limited in extent.

Focal or generalized vitiligo exhibits a progressive spread, involving small or large portions of the skin anywhere on the body without following any predictable patterns. Due to the interchangeable and arbitrary nature of vitiligo lesions, their classification may change over time.

A helpful classification system based on the activity of the disorder is V1, V2, and V3, which aids in accurately assessing the patient's condition and deciding the appropriate line of therapy **(Table 2)**.

Additionally, cases of vitiligo can be further classified based on the distribution and severity of the disease, which can facilitate effective management and treatment planning.

TABLE 1: Classification of vitiligo depending on the activity of disease (Behl).

Stages of vitiligo	Clinical features
Active	• New lesions developing • Lesions increasing in size • Border ill-defined
Quiescent	• No lesions developing • Lesions stationary in size • Border hyperpigmented and well defined
Improving	• Lesions decreasing in size • No new lesions developing • Border defined and signs of spontaneous repigmentation (follicular and peripheral)
Zosteriformis	• Unilateral distribution of lesions • Lesions increasing in size • Borders ill-defined

TABLE 2: Classification of vitiligo based on activity of the disorder.

Stage of vitiligo	Treatment recommended
V1	Conservative therapy with vitamins. Copper, protein supplements, may be antihistamines and steroids
V2	Psoralens, Ammi majus and conservations
V3	Surgical intervention in cases with no further improvement

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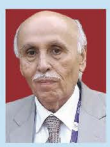
A Handbook of
VITILIGO & Color Atlas

Salient Features

- This edition of the book of vitiligo has undergone comprehensive revisions to all chapters, with updates and expanded content providing the reader with a more comprehensive and in-depth coverage of treatment modalities of vitiligo with a section "Color Atlas"
- Drugs derived from living organisms discussed throughout
- Presents an approach and evaluation of vitiligo and pigmentation disorders in general
- Deals with various etiological causes and modalities of treatment and management of the disease
- Provides extensive data on the basic science surrounding leukoderma, pigmentation, and pigmentary disorders
- Provides new treatments—both medical and cosmetic surgical modalities—in large
- Includes information about recent therapy, use of lasers, surgery, tattooing, and other treatments
- *"A Handbook of Vitiligo and Color Atlas"* is a companion for busy practicing dermatologists, residents, and family physicians.



SK Punshi MBBS DDV FIMS FDS is a Senior Consultant and Dermatologist in Amravati, Maharashtra, India. He is an acclaimed and undeniable clinical research worker, especially in the field of vitiligo. His work on leukoderma is quoted in various books and journals of dermatology. He is the first skin specialist in the world to introduce placental extract therapy and herbomineral therapy in vitiligo.

Formerly, he was the President of Indian Medical Association (Amravati), Joint Secretary of Indian Medical Association (Maharashtra), President of Indian Association of Dermatologists, Venereologists and Leprologists (Maharashtra), Vice President of Dermatological Society of Vidarbha, and Vice President of All India Association of Dermatologists, Venereologists, and Leprologists.

He received many awards like All India Award by Skin Institute and School of Dermatology, New Delhi, for his research thesis on placental extract in vitiligo; Ambady Oration Award in IADVL Conference (Jabalpur) on Vitiligo: Yesterday and Today (1986); and Lions International Club Award for his contribution in leprosy eradication. He attended a number of national and international conferences and chaired many scientific sessions at several regional, state, national, and international conferences in his 45–50 years of career in dermatology. Dr Punshi has written many articles in literary journals and presented many regional and national papers and authored many books on health awareness.

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