Vitiligo: How many types?

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Vitiligo is one of the most common cutaneous disorders. However, evidence-based treatment guidelines were missing.

This textbook:
- Fills a void in the literature.
- Is written by the most authoritative experts in the field.
- Includes therapy recommendations based on new evidence-based guidelines.
- Demonstrates case studies with clinical pictures before and after the treatment in order to illustrate the treatment success.

It defines and gives a complete overview of the disease, both regarding the classification of differential diagnosis as well as the treatment. Thus, it will be a valuable resource for all physicians who are seeing patients with this stigmatizing disease.
Vitiligo

- Affects around 0.5 of the population worldwide
- Etiology poorly understood
- Non-disease status
- Stigma and perceived severity
- Near orphan disease (especially for drug development)

Picardo & Taïeb, Vitiligo, Springer 2010
Vitiligo, a puzzling SKIN disease

Picardo & Taïeb, Springer, 2010
From the standpoint of genetic susceptibility, the TYR Arg402Gln polymorphism represents an inverse relationship between NSV and malignant melanoma.

*Spritz, Genomic Med, 2010*
VETF: NSV Definition

- Acquired
- Chronic
- Pigmentation disorder
- White patches
- Often symmetrical
- Substantial loss of functioning epidermal and/or hair follicle melanocytes
- Patches usually increasing in size with time

Ghent 2003

PCMR 2007 VETF consensus paper
Initial assessment and follow-up

PCMR 2007 VETF consensus paper
NS Vitiligo Excludes

- Piebaldism & other heritable circumscribed hypomelanoses incl tuberous sclerosis
- Post inflammatory depigmentation (incl MF)
- Post infectious depigmentation: pityriasis versicolor, leprosy
- Post traumatic leucoderma
- Melanoma-associated leucoderma
- Melasma
- Drug induced depigmentation (topical and systemic)
Diagnostic Quizzes
Diagnostic Quizzes

PSORIASIS

NEVUS DEPIGMENTOSUS
Piebaldism
Diagnostic Quizzes
Diagnostic Quizzes

Lichen sclerosus

Vitiligo following LS
Diagnostic Quizzes
Diagnostic Quizzes
Diagnostic Quizzes

Vitiligo + type 1 Diabetes
Diagnostic Quizzes
Diagnostic Quizzes

Vitiligo + atopic dermatitis
NSV-SV: distinct disorders?

Segmental Vitiligo
- Often begins in childhood
- Has rapid onset and stabilizes
- Involves hair compartment soon after onset
- Is usually not accompanied by other autoimmune diseases
- Often occurs on the face

Is usually responsive to autologous grafting, with stable repigmentation

Can be difficult to distinguish from nevus depigmentosus, especially in cases with early onset

Nonsegmental Vitiligo
- Can begin in childhood, but later onset is more common
- Is progressive, with flare-ups
- Involves hair compartment in later stages
- Is often associated with personal or family history of autoimmunity
- Commonly occurs at sites sensitive to pressure and friction and prone to trauma

Frequently relapses in situ after autologous grafting

Taïeb & Picardo, NEJM 2009
Type of melanocytic target
Multivariate analysis of factors associated with early onset segmental and non-segmental Vitiligo: a prospective observational study of 213 patients


BJD 2011
Usually SV is considered as rather dermatomal.
SV+NSV: Mixed Vitiligo

Gauthier et al, PCR 2003
NSV+ SV: more common than recognized?

Taïeb et al, PCMR 2008
Segmental vitiligo associated with generalized vitiligo (mixed vitiligo): A retrospective case series of 19 patients


Ezzedine et al, JAAD 2011
VITILIGO GLOBAL ISSUES
CONSENSUS
CONFERENCE
1. SEOUL 22nd WCD
2. BORDEAUX 21st IPCC
« Umbrella » term for nonsegmental vitiligo

- Non-segmental vitiligo
- Common vitiligo
- General vitiligo
- Bilateral vitiligo
- Generalized vitiligo
- Vitiligo +++
Other elements of consensus

- **Segmental vitiligo**: segmental pattern should be clinically clear (*vs* Focal); no reference to type of pattern (no consensus)

- **Mixed vitiligo** (MV) to be included in the classification as a variant of vitiligo (formerly NSV) 1st step: SV, 2nd step V (NSV)

- **Occupational vitiligo**: Premature to be included, not etiology-based classification

- **Unclassified**: focal, pure mucosal

- **Rare possible variants** defined in glossary.
# VITILIGO CLASSIFICATION

Bordeaux VGICC 2011

<table>
<thead>
<tr>
<th>Subtypes</th>
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</thead>
<tbody>
<tr>
<td>Vitiligo/NSV</td>
</tr>
<tr>
<td>Acrofacial</td>
</tr>
<tr>
<td>Mucosal (more than one mucosal site)</td>
</tr>
<tr>
<td>Generalized</td>
</tr>
<tr>
<td>Universal</td>
</tr>
<tr>
<td>Mixed (associated with SV)</td>
</tr>
<tr>
<td>Rare variants</td>
</tr>
<tr>
<td>Uni-, bi-, or plurisegmental</td>
</tr>
<tr>
<td>Focal</td>
</tr>
<tr>
<td>Mucosal (one site in isolation)</td>
</tr>
<tr>
<td>Segmental Vitiligo</td>
</tr>
<tr>
<td>Undetermined/ unclassified Vitiligo</td>
</tr>
</tbody>
</table>

**NSV, non-segmental vitiligo; SV, segmental vitiligo; VGICC, Vitiligo Global Issues Consensus Conference.**

Ezzedine et al, PCMR 2012
V. punctae
The term « autoimmune vitiligo » should not be included in the classification.

Vitiligo (NSV) seems driven by immune-mediated mild inflammatory mechanisms in most cases, but the relation of local to general autoimmunity is not clear.

Ezzedine et al, PCMR 2012
No objective clinical evidence of skin inflammation in common vitiligo

- No redness or edema in common vitiligo
- Significance of « Pruritus » item in VETF form
- Most evidence comes from histopathology, but:
  - Stage/progression related?
  - Universal or uncommon/rare feature?
  - Shared between SV and NSV?
NSV: microinflammation nearly constant in progressing borders

Photographs B Vergier, CHU de Bordeaux
SV: evidence of an inflammatory stage in early lesions

Patient of Dr Attili, India

Ezzedine et al in: Vitiligo, Picardo & Taïeb, 2010
VETF Position paper on KP

Koebner

Type 1
History

Type 2
Clinically

Type 3
Experimentally induced

Depigmentation after trauma during last year?

Type of trauma:
1. Physical (wound, cuts, scratching)
2. Mechanical (friction)
3. Chemical/thermal (burns)
4. Allergic (contact dermatitis or irritant reactions (vaccination, tattoos…)
5. Chronic pressure
6. Inflammatory dermatoses
7. Therapeutics (radiotherapy, phototherapy, …)

A. Depigmentation corresponding either to areas of repeated pressure or friction (elbows and knees) or areas of chronic friction related to cloths/accessory.

B. Depigmentation clearly induced by trauma (linear, punctiform, crenate)

Eliciting trauma
-I: repeated pressure/friction
-II: Superficial (epidermal) trauma
-III: Dermo-epidermal trauma

Underwear print pattern suggestive of previous injury

Van Geel et al, PCMR 2011
Koebner’s phenomenon: link trauma-inflammation?

Taieb & Picardo NEJM 2009

K-VSCOR for scoring Koebner’s phenomenon

Diallo et al, PCMR 2013

K-VSCOR (0-100). Seven variables independently associated with the presence of KP: disease duration of more than 3 years, forehead + scalp areas, eyelids, wrists, genital + belt areas, knees and tibial crests.
Identification of 2 phenotypes of non-segmental vitiligo: a latent class analysis of a 717-patient series

K. Ezzedine, A. Le Thuaut, T. Jouary, F. Ballanger, A. Taieb, S. Bastuji-Garin

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Public Health department and Université Paris Est Créteil (UPEC), LIC EA4393, Créteil, France
LCA: summary of results

• 2 subtypes of vitiligo, major distinct features:
  – prepubertal:
    • trunk and limbs locations; halo nevi, family history of hair graying, family history of vitiligo/NSV
    • atopic dermatitis and family history of other autoimmune disorders
  – postpubertal:
    • “pure” acrofacial pattern without lesions on trunk and limbs more common
    • Role of major stresses seems more important

Ezzedine et al, IID Edinburgh 2013
Vitiligo: How Many Types?

- SV: developmental pattern ++
- Vitiligo/NSV: 2 types pre and postpubertal? TBC
- Koebner’s linked to acceleration (inflammatory) phase TBC in SV
- Acceleration phase always immune-mediated inflammation: TBC in SV
- « Continuum » view SV-V more in line with current data.
- Importance for pathophysiology and therapy: combined targets: cutaneous inflammation/melanocyte regeneration
35th COURSE OF PEDIATRIC DERMATOLOGY and 3rd European course, ARCACHON, FRANCE
22-24 April 2014
http://www.dermatobordeaux.fr