

Vitiligo: How many types?



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Vitiligo

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.

Picardo
Taïeb
Editors
Vitiligo



Vitiligo



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> springer.com

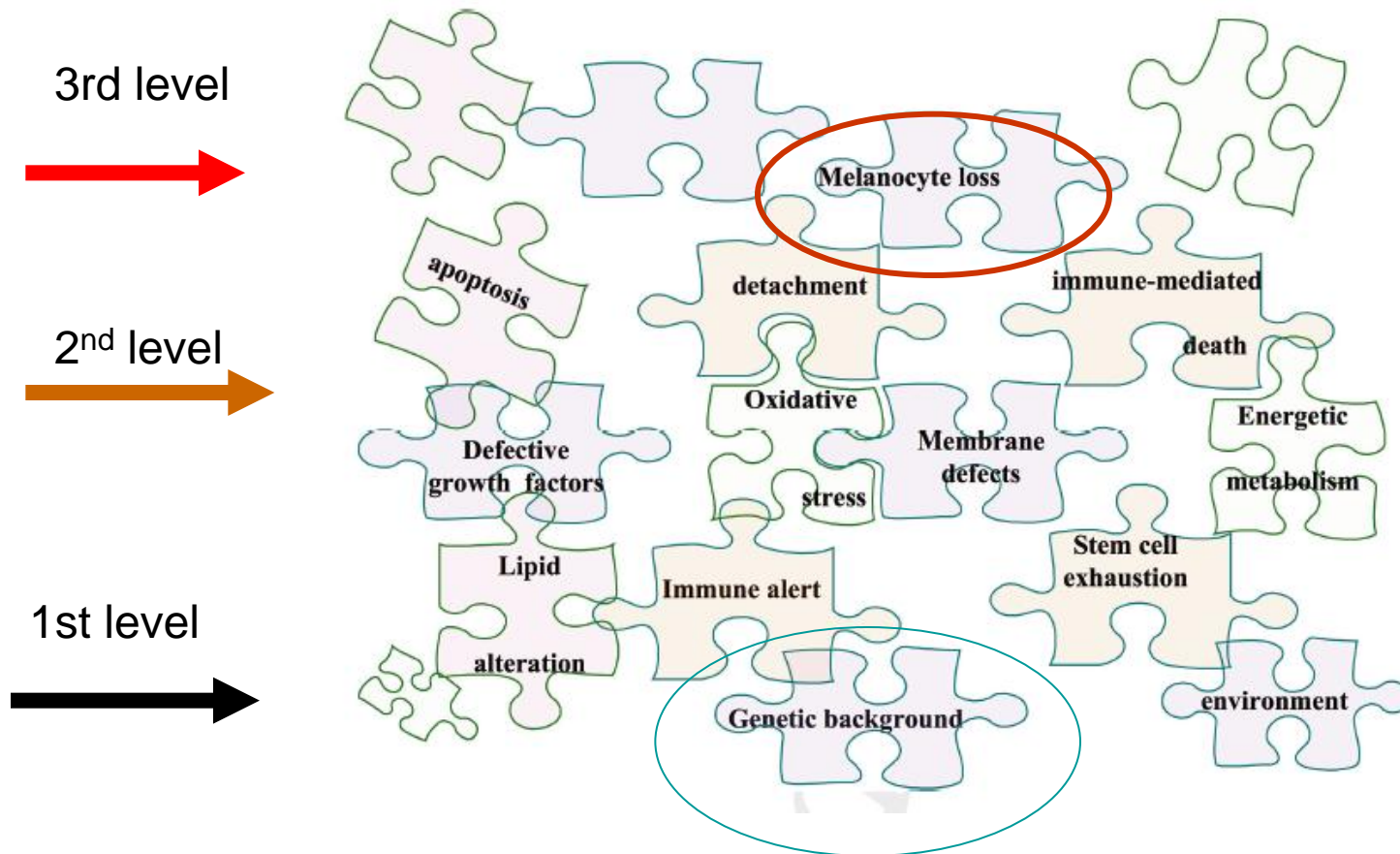
 Springer

RELEASED 2010

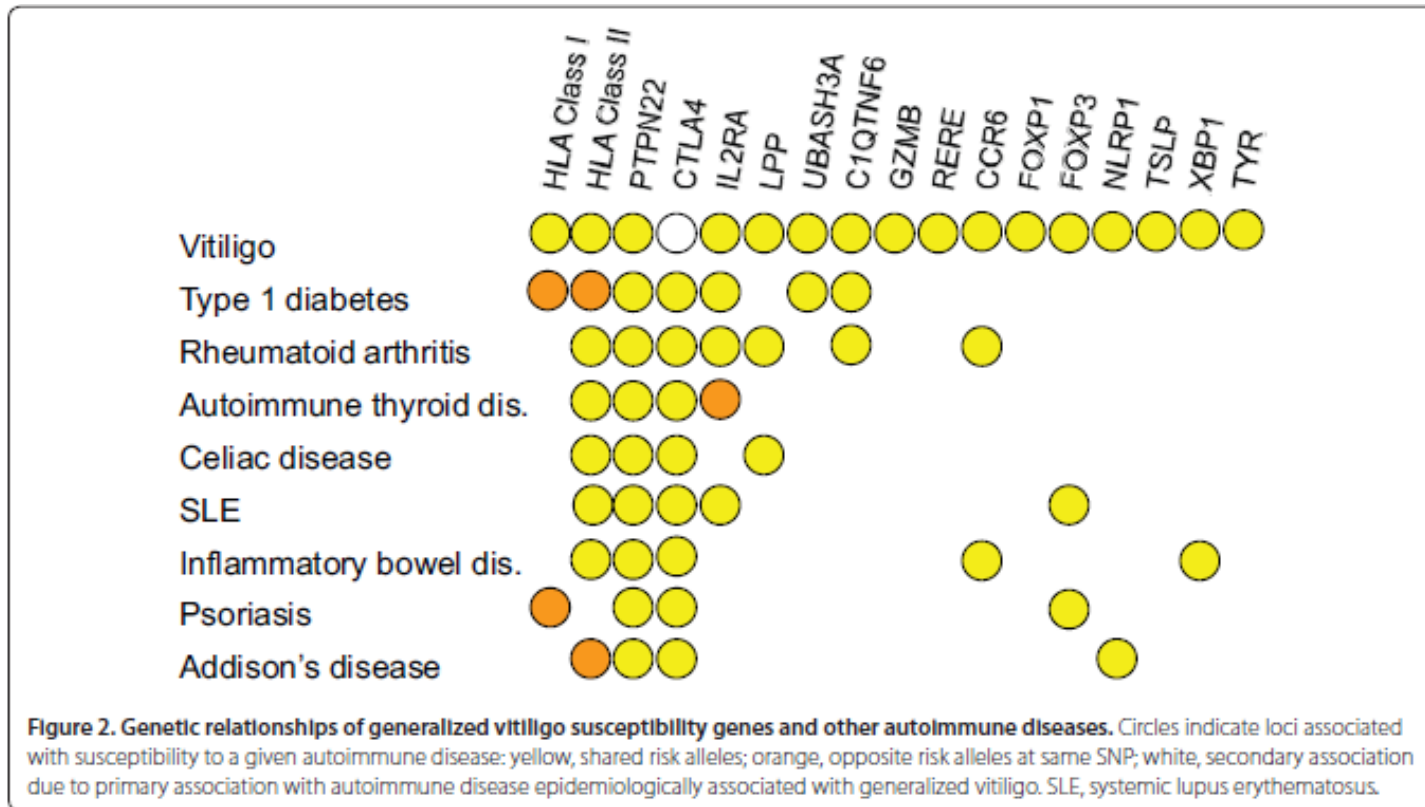
Vitiligo

- 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)

Vitiligo, a puzzling SKIN disease



Pangenomic studies (GWAS)



From the standpoint of genetic susceptibility, the *TYR* Arg402Gln polymorphism represents an inverse relationship between NSV and malignant melanoma

VETF: NSV Definition

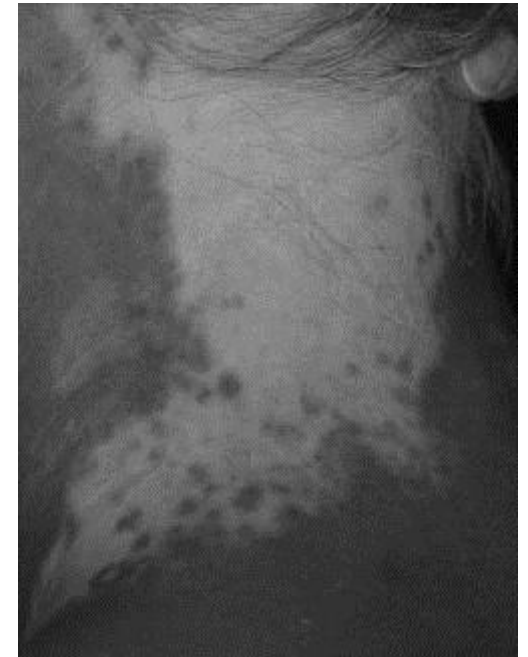


- Acquired
- Chronic
- Pigmentation disorder
- White patches
- Often symmetrical
- Substantial loss of functioning epidermal and /or hair follicle melanocytes
- Patches usually increasing in size with time

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



PMH



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



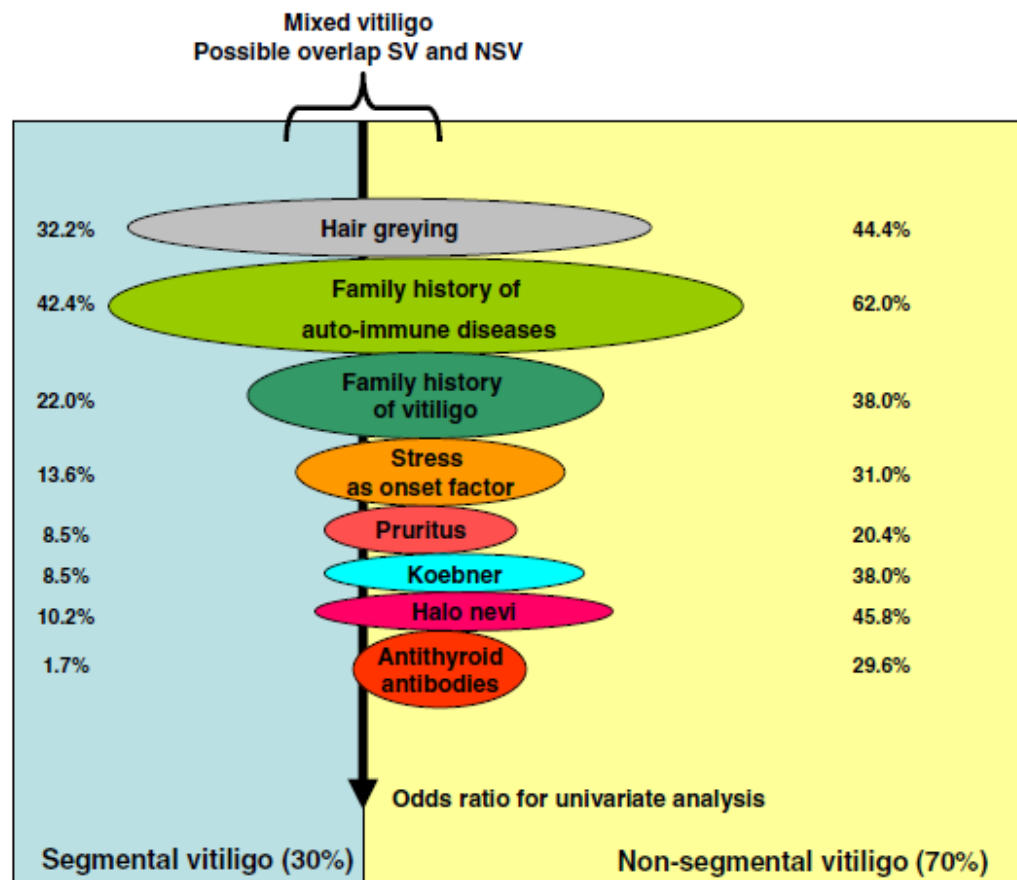
Multivariate analysis of factors associated with early onset segmental and non-segmental

Vitiligo: a prospective observational study of 213 patients

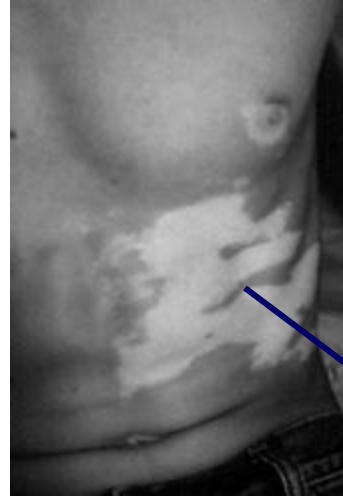
K. Ezzedine*, A. Diallo, C. Léauté-Labrèze, D. Mossalayi, Y. Gauthier, S. Bouchtnei, M.

Cario-André, J. Seneschal, F. Boralevi, T. Jouary, A. Taieb

BJD 2011



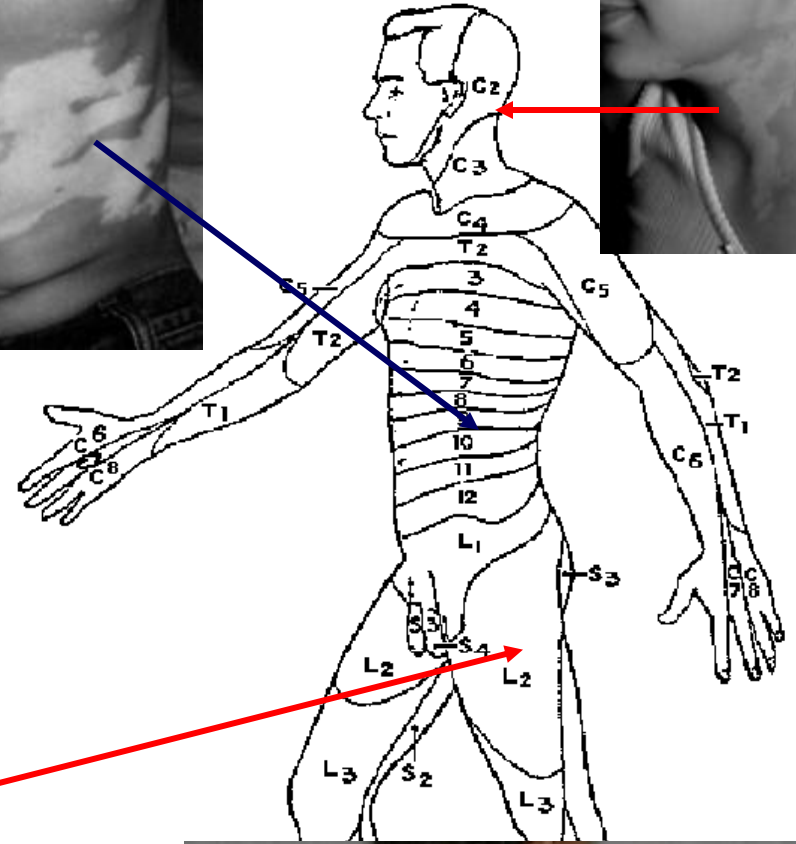
Usually SV is considered as rather dermatomal



C2 ?



T6-10?



L2 ?

Taïeb et al, PCMR, 2012

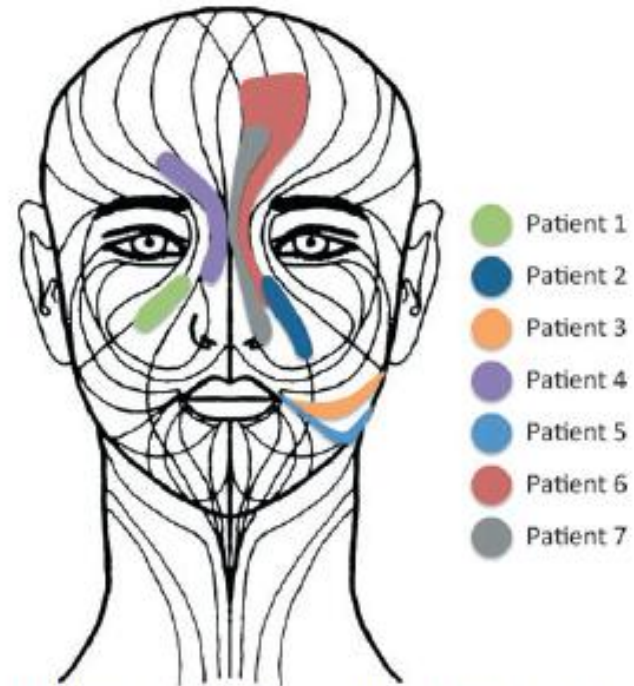


Figure 1. A schematic representation of the lines of Blaschko on the face with skin lesions from 7 patients diagnosed with lichen striatus. Image adapted from Happle and Assim, 2001 (15).

Taieb A, el Youbi A, Grosshans E, Maleville J. Lichen striatus: a Blaschko linear acquired inflammatory skin eruption. *J Am Acad Dermatol*. 1991 Oct;25(4):637-42.

SV+NSV: Mixed Vitiligo



Gauthier et al, PCR 2003

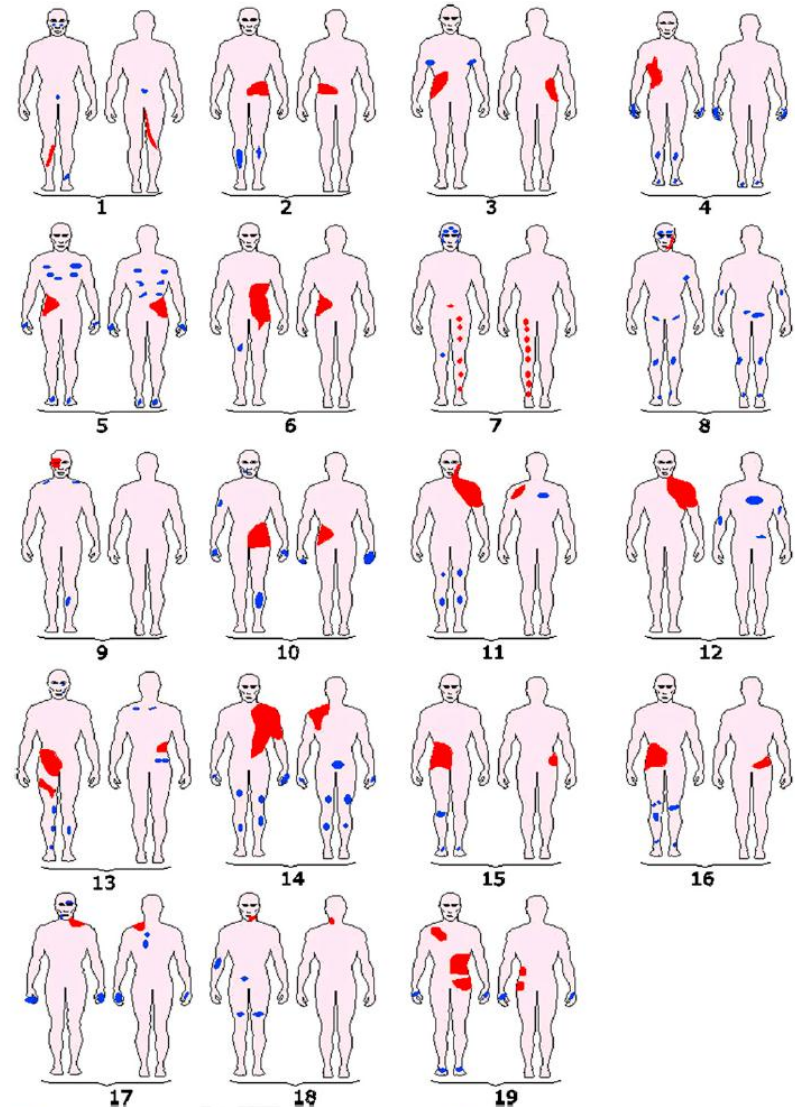
NSV+ SV: more common than recognized?



Taïeb et al, PCMR 2008

Segmental vitiligo associated with generalized vitiligo (mixed vitiligo): A retrospective case series of 19 patients

Happle R. Superimposed segmental manifestation of polygenic skin disorders. *J Am Acad Dermatol* 2007;57:690-9.



Ezzedine et al, *JAAD* 2011

Fig 1. Pattern of segmental (red) and nonsegmental (blue) involvement in 19 patients with mixed vitiligo.

VITILIGO GLOBAL ISSUES CONSENSUS CONFERENCE

1. SEOUL 22nd WCD
2. BORDEAUX 21st IPCC





Bordeaux VGICC 21 Sept 2011

« 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) 1ststep: 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 3. Bordeaux VGICC classification and consensus nomenclature

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

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



V Minor



Follicular V



V. punctue

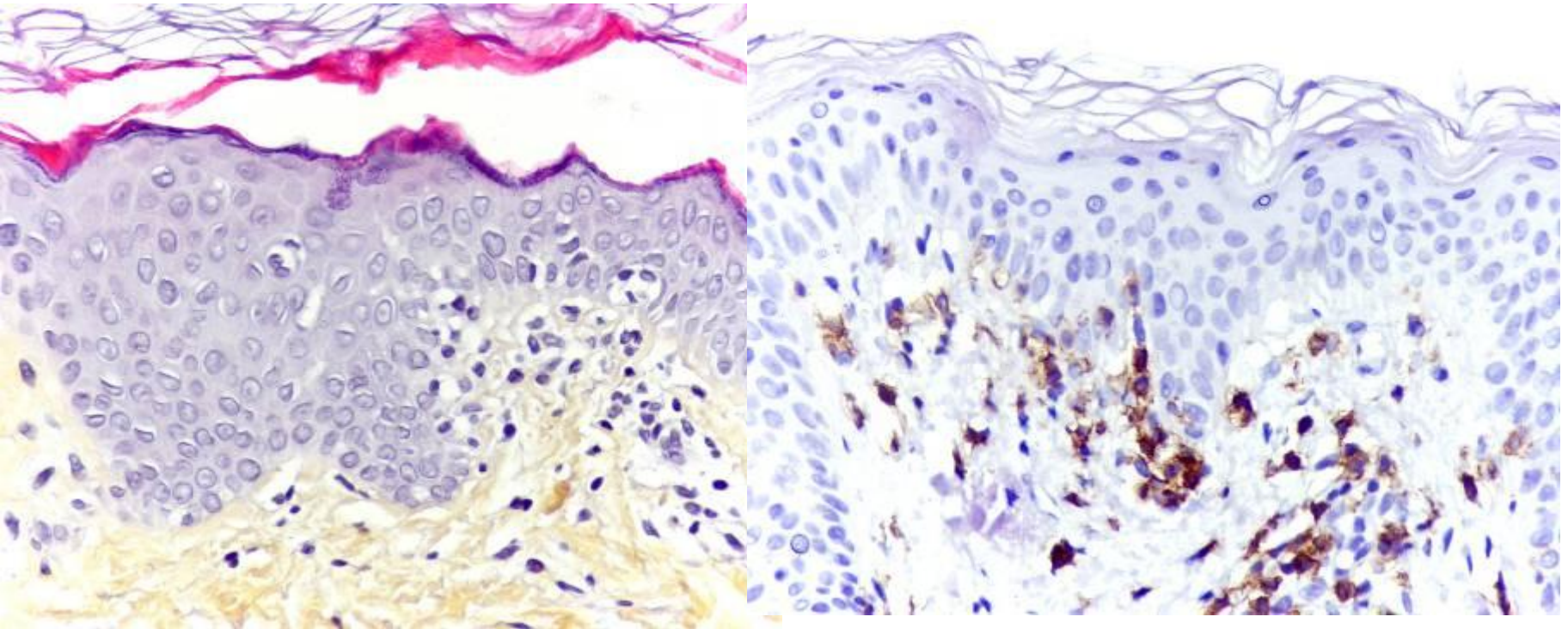
Statements at Consensus conference

- 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.*

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



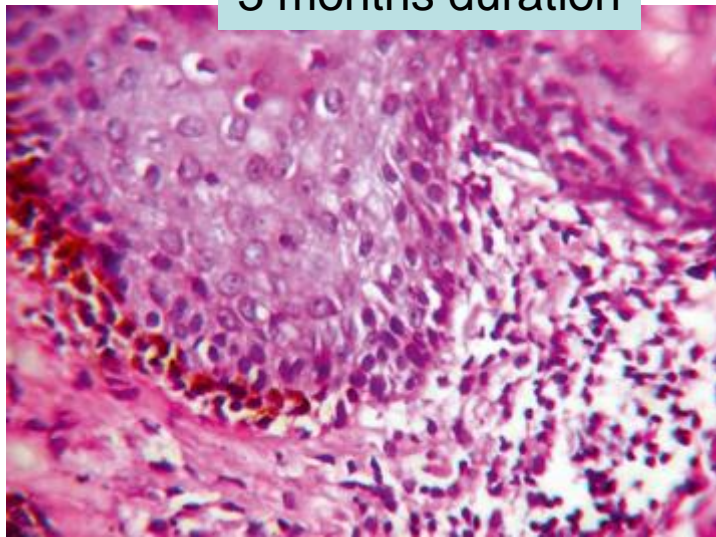
CD8

SV: evidence of an inflammatory stage in early lesions



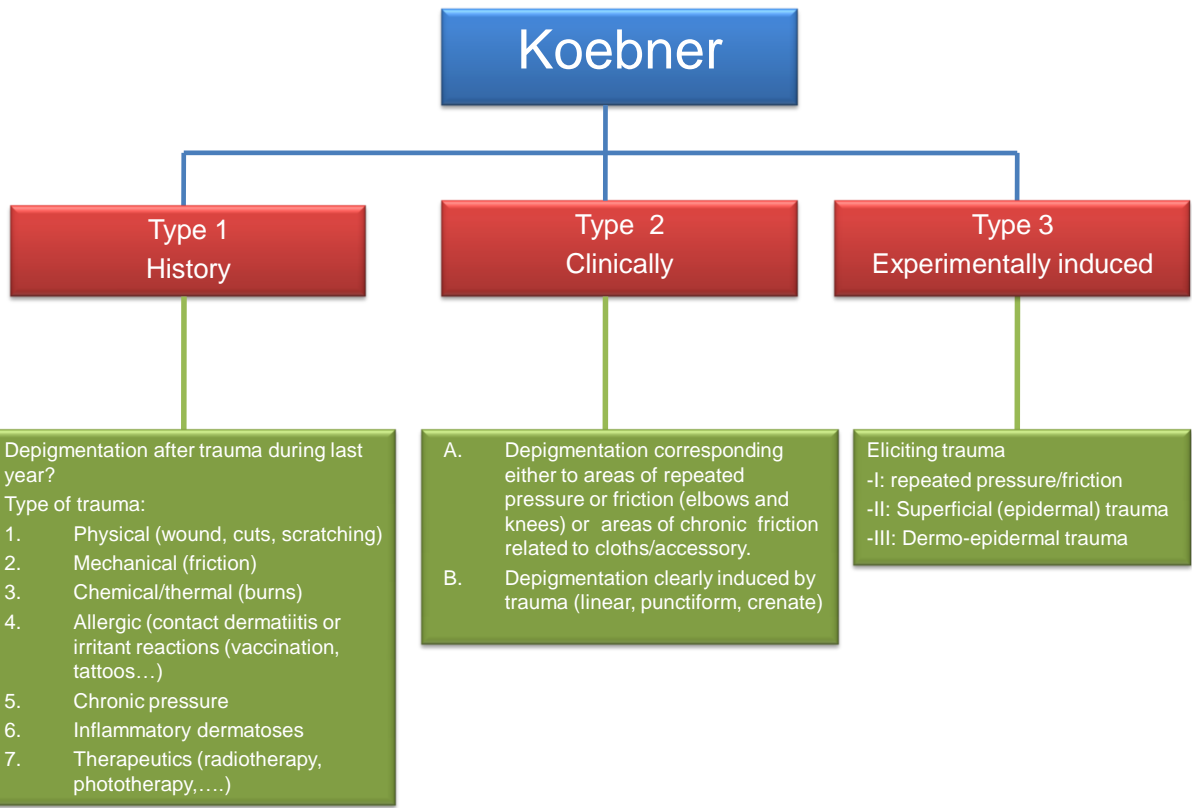
3 months duration

+ 4 months



Patient of Dr Attili, India

VETF Position paper on KP



2A
Underwear print pattern

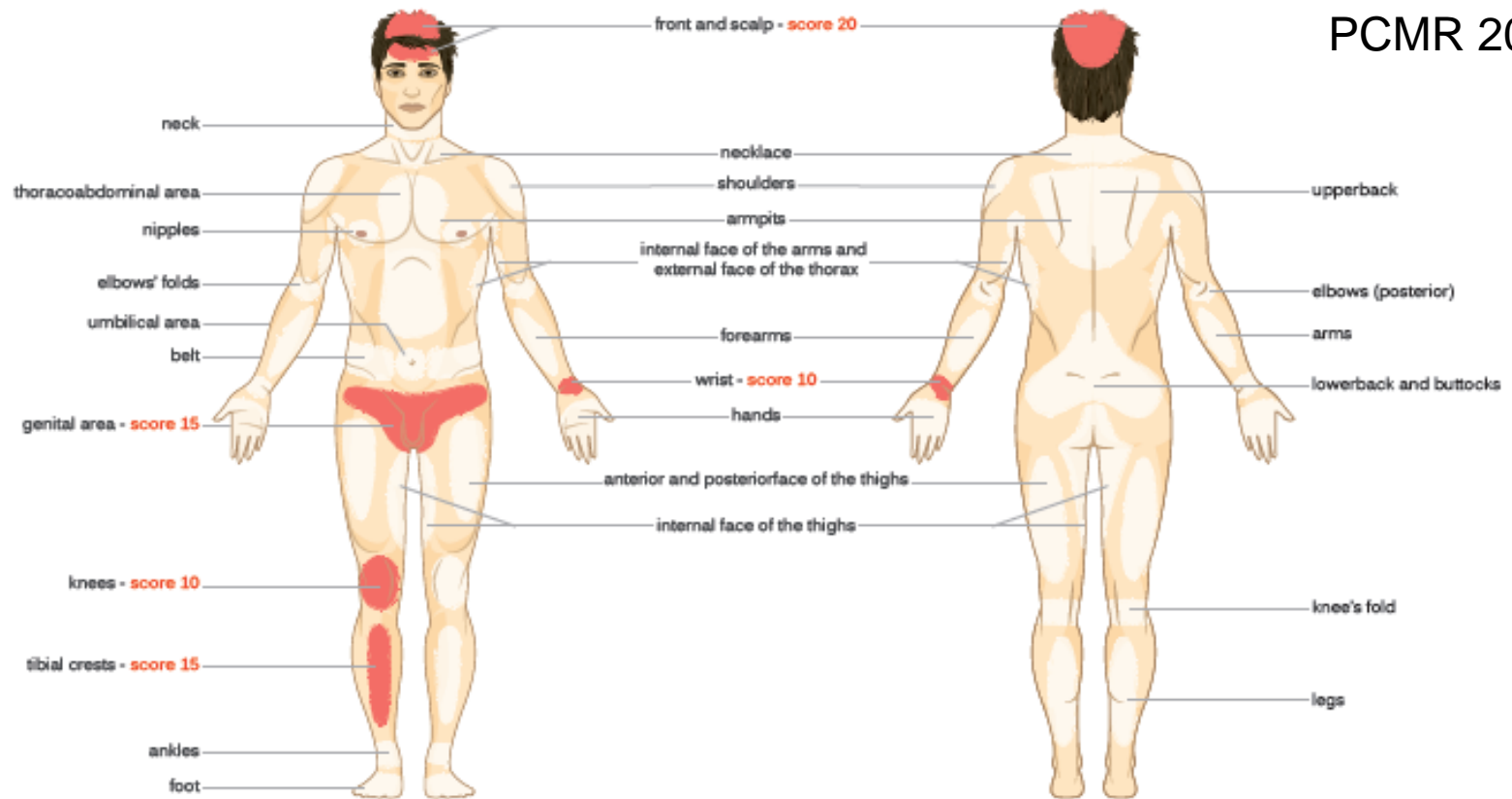


2B

suggestive of previous injury

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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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

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



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