VII Masterclass on
Vitiligo and **Pigmentary** Disorders

21-23 November 2014
İstanbul Florence Nightingale Hospital İstanbul - Turkey

Vitiligo
Clinical Presentations

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Agenda

- Clinical features of vitiligo
- Other cutaneous and systemic abnormalities in vitiligo
- Childhood vitiligo
- Course of vitiligo
- Diagnosis
- Differential diagnosis

Vitiligo

- a common, acquired depigmentation disorder of the skin and mucous membranes
- the prevalence is around 1% of the population
- M=F
- Half of the patients develop the disease before 20 years of age

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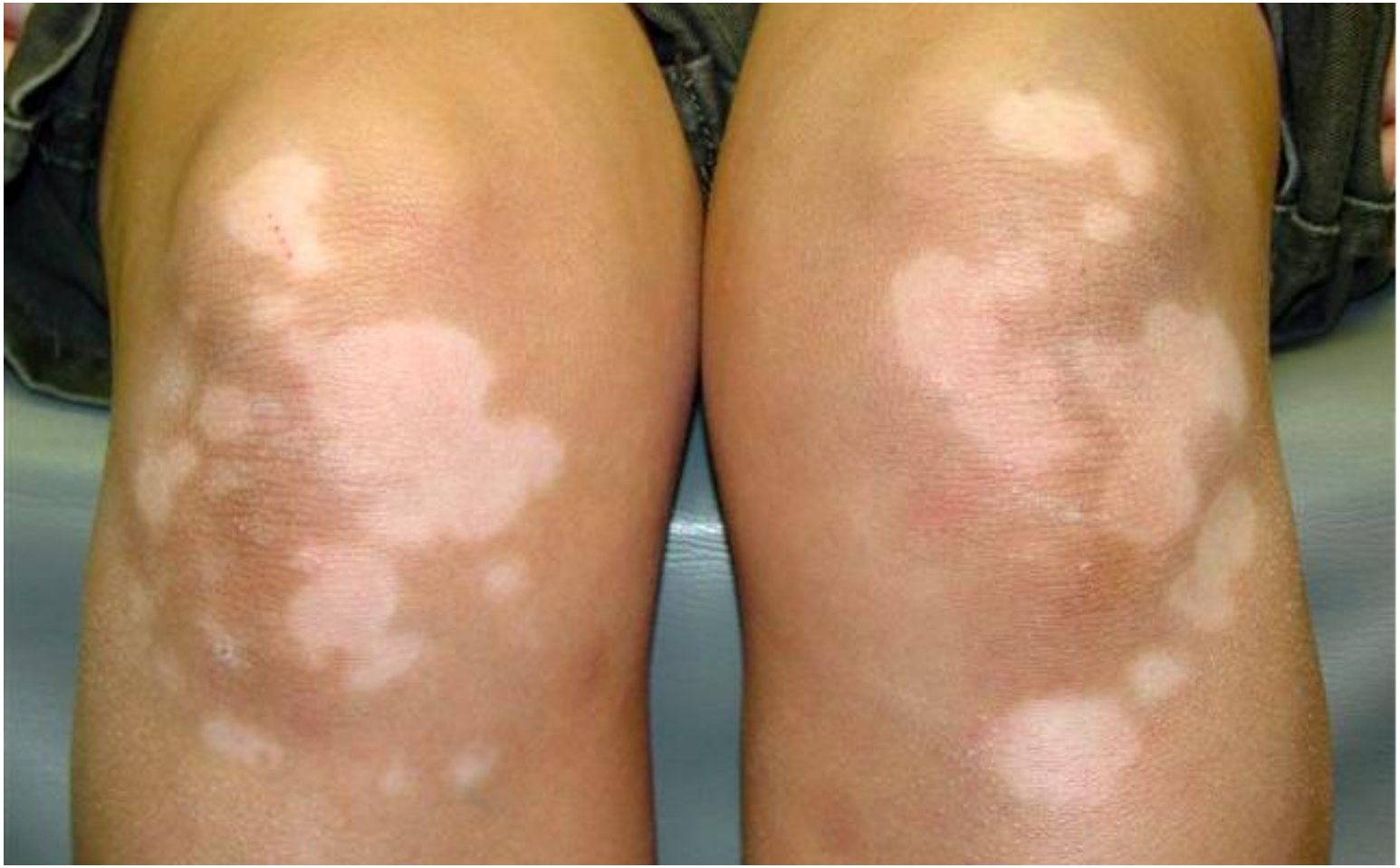
Clinical progress of Vitiligo

- Vitiligo usually begins in sun-exposed areas
- Severe sunburn, pregnancy, skin trauma, and/or emotional stress may be precede onset.
- Vitiligo generally slowly progressive, either by centrifugal expansion of current lesions or appereance of new lesions

Clinical progress of Vitiligo

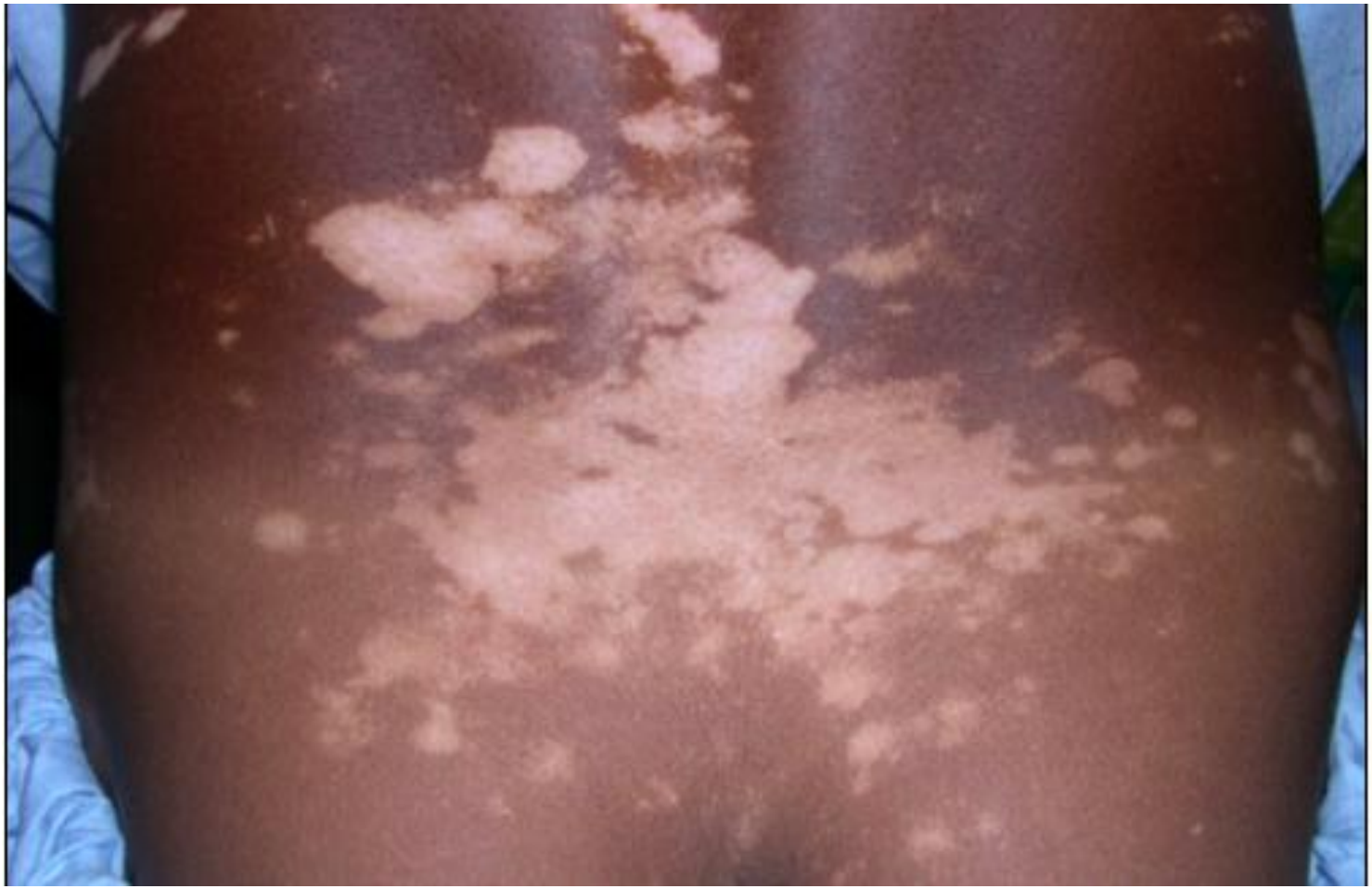
- Vitiligo can occur anywhere on the body
- On face, it favors around the eyes and periorificial areas
- On extremities, it favors the elbow, knees, digits and flexor wrists
- Koebnerization is typical.
- Koebner phenomenon is an important parameter to assess and predict the clinical profile and progress of vitiligo.

The typical vitiligo lesion



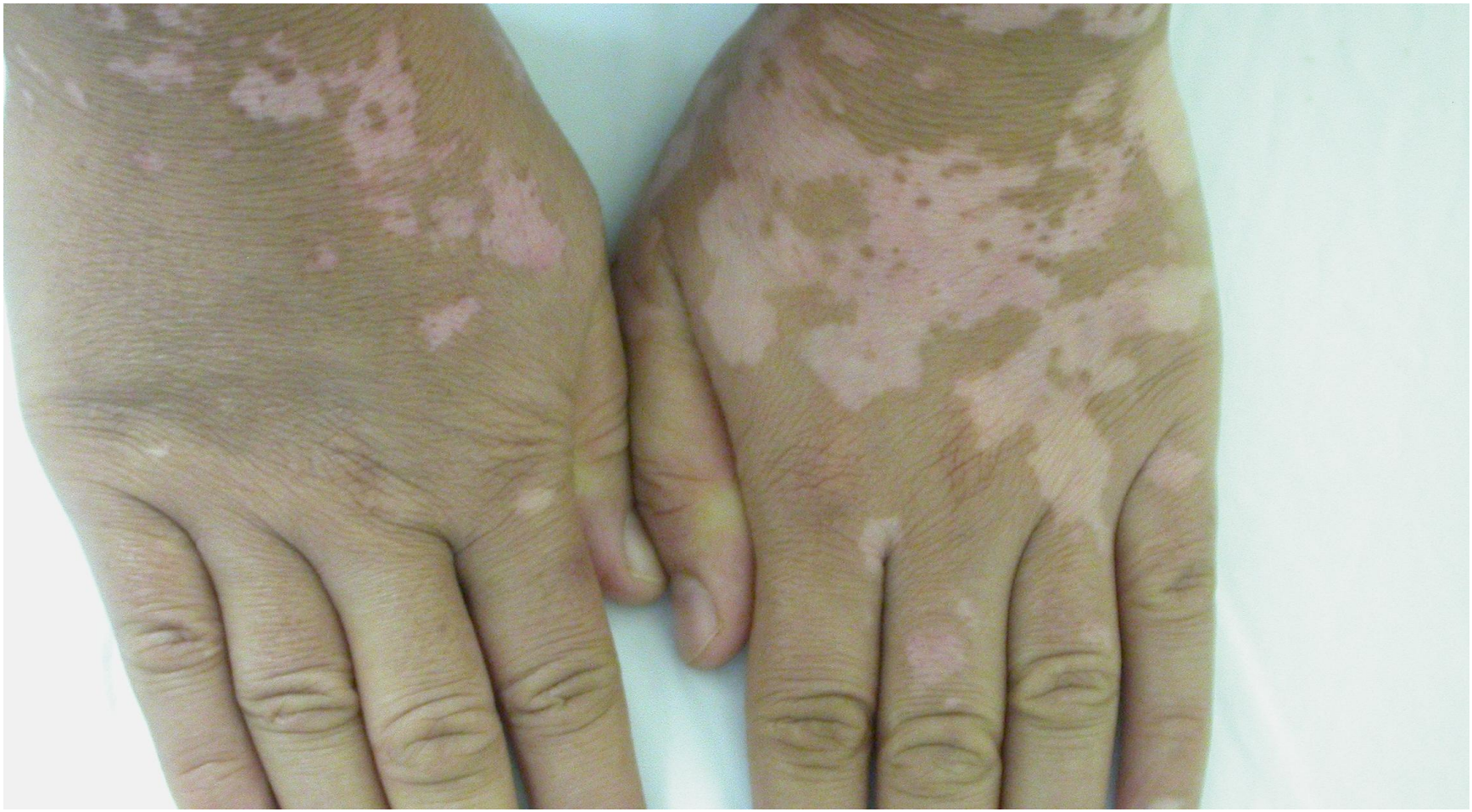
Variations on the typical vitiligo macule

Trichrome vitiligo



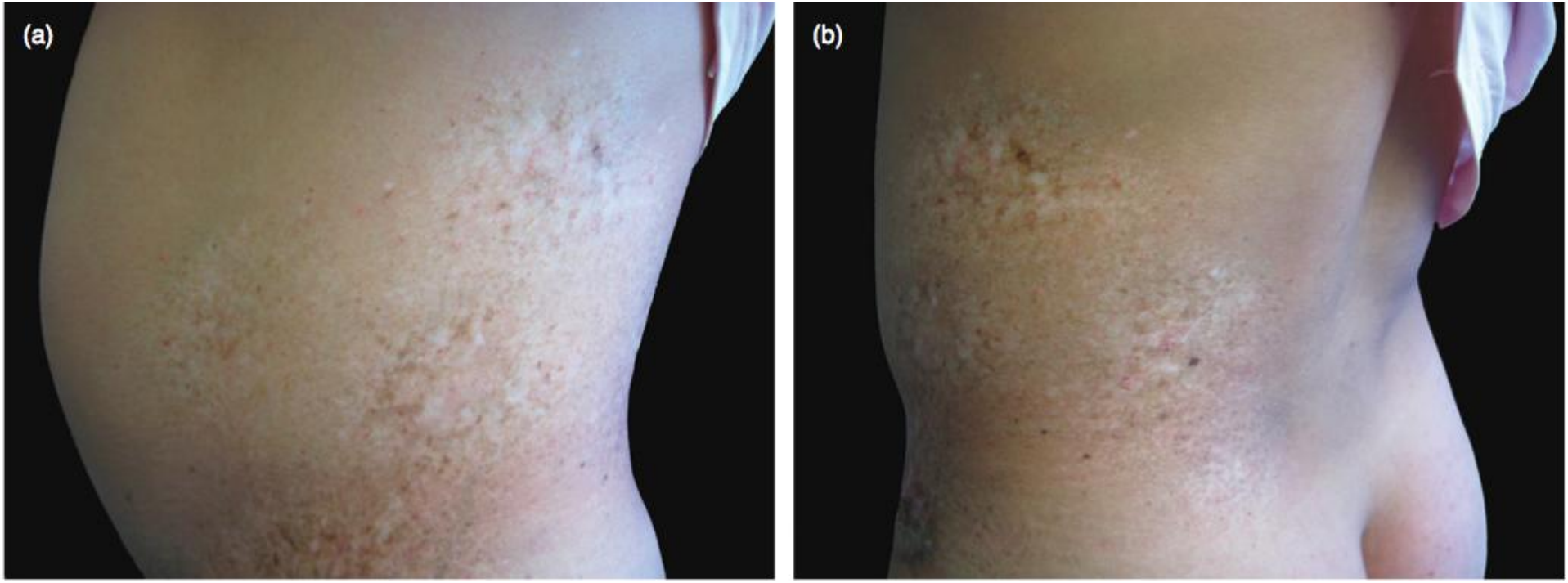
An uniform tan coloration zone between normal and depigmented skin

Quadrichrome vitiligo



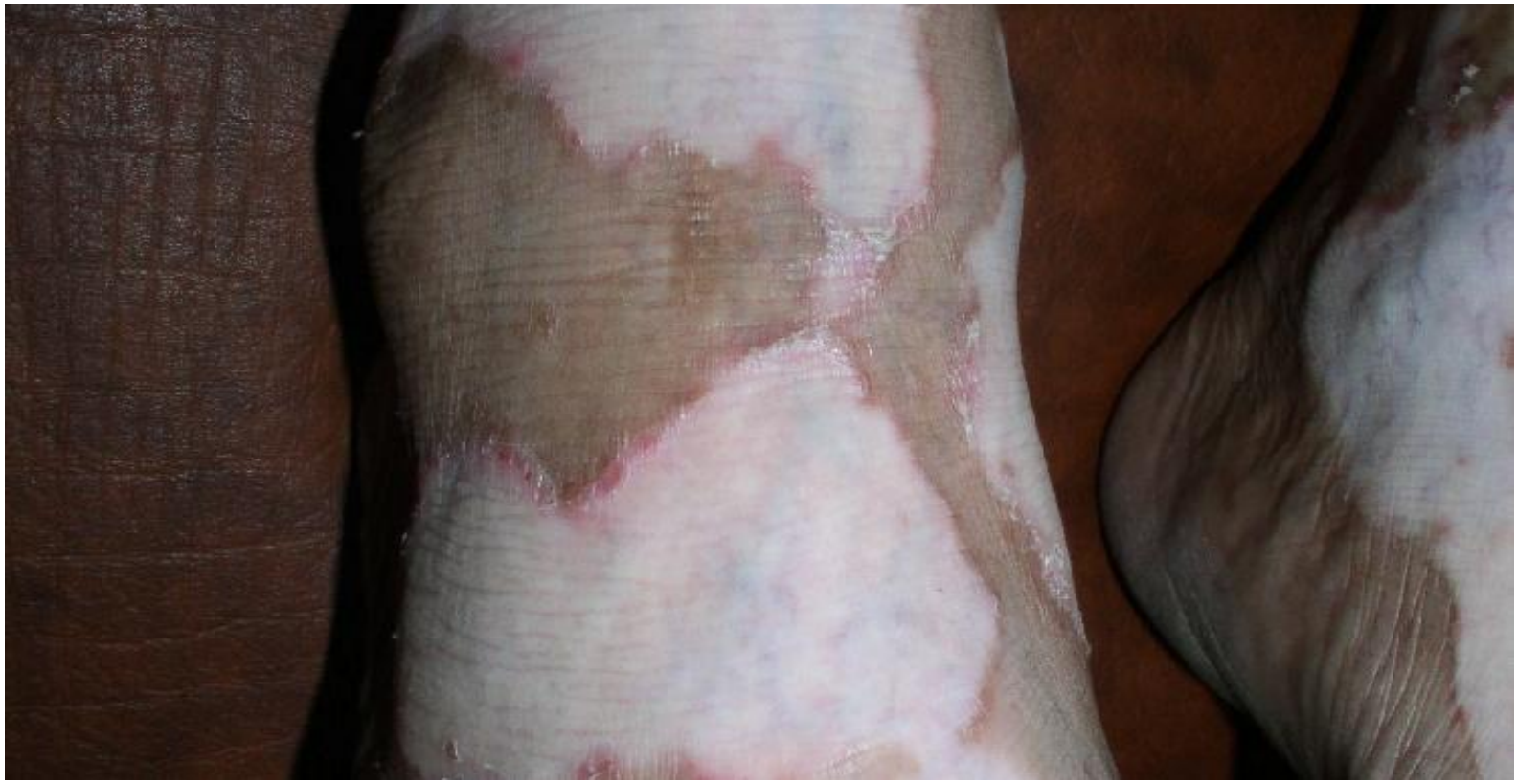
An additional macular perifollicular hyperpigmentation of repigmented areas

Pentachrome vitiligo



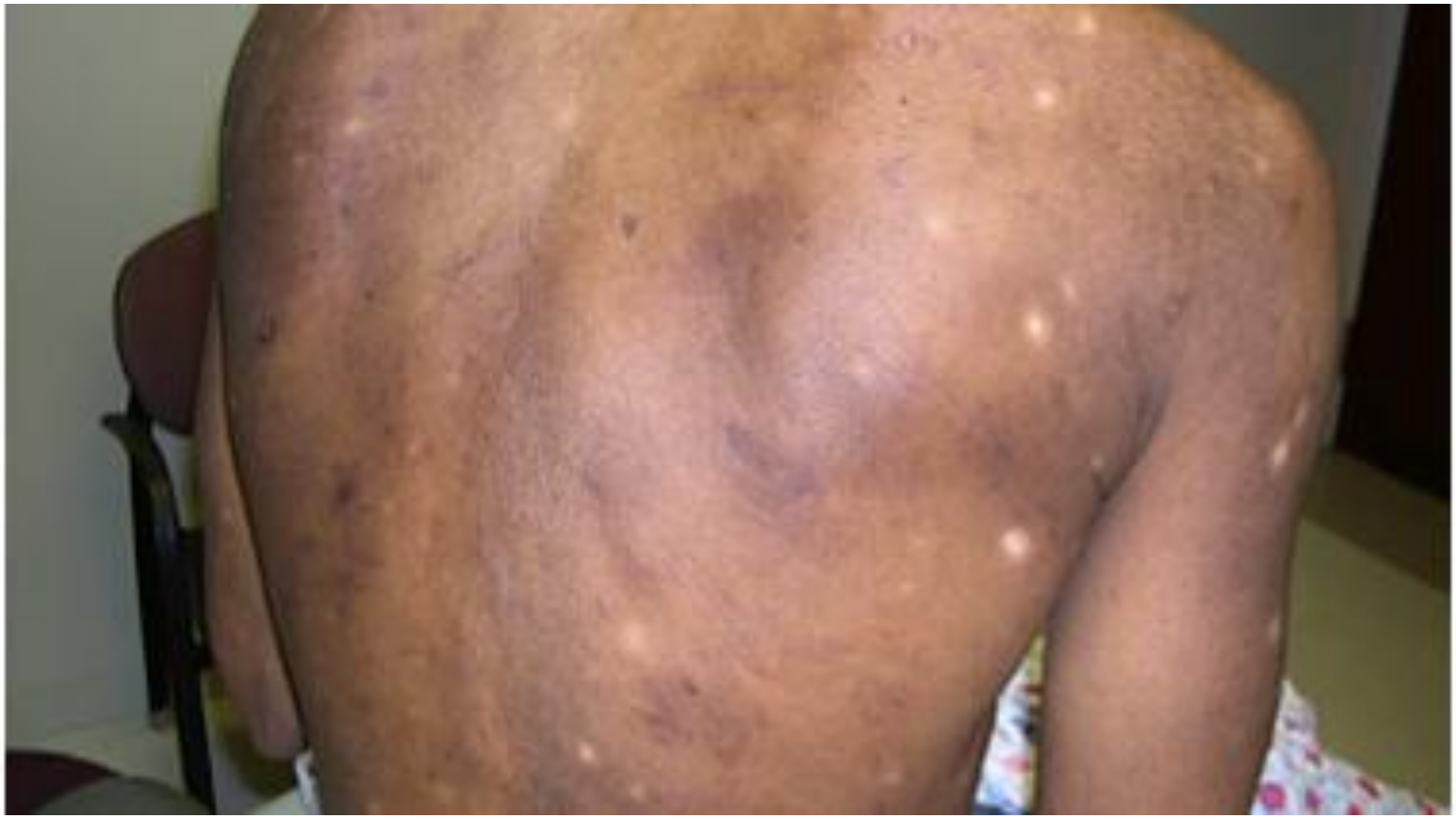
- ✓ Five shades of colour: white, tan, brown, blue-gray, normal
- ✓ Blue vitiligo corresponds to vitiligo macules in sites of postinflammory hypermelanosis

Inflammatory vitiligo



- ✓ Erythema at the margins of depigmented macules
- ✓ A red and raised border
- ✓ Mild pruritus may be present

Vitiligo ponctue



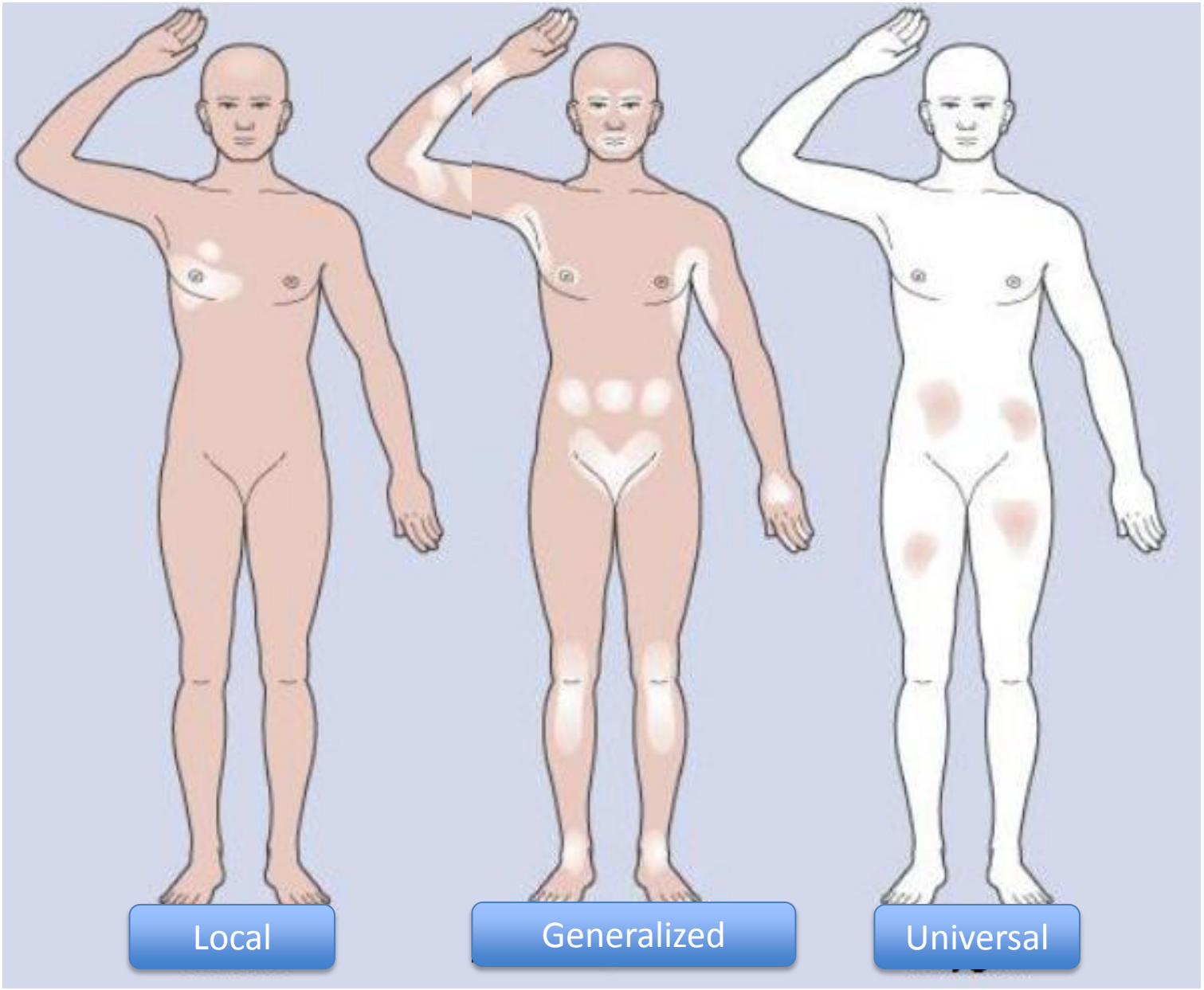
Discrete, confetti-like amelanotic macules on normal or hyperpigmented skin

Clinical classification of Vitiligo

Clinical classification of Vitiligo

1. Localized
2. Generalized
3. Universal

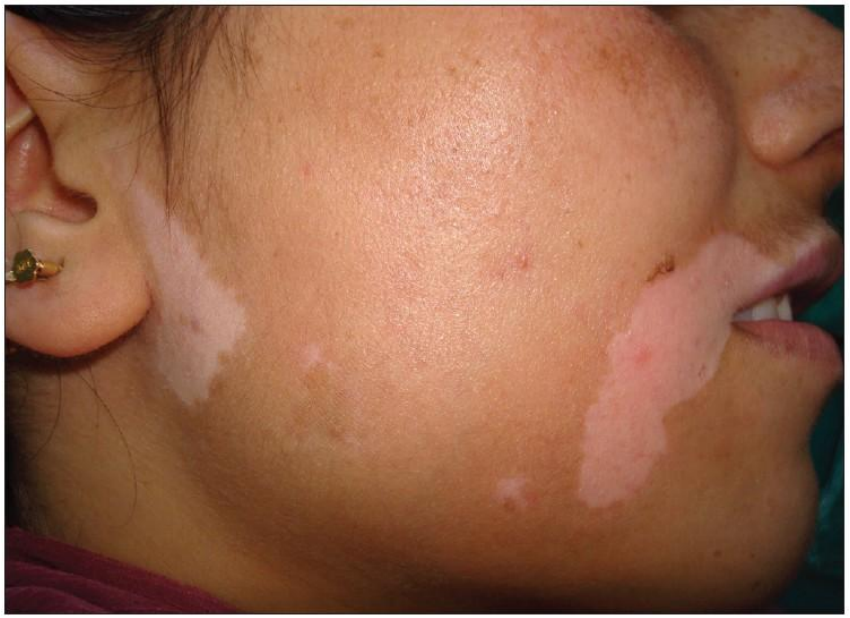
Distribution pattern of amelanotic skin lesions in vitiligo



Localized Vitiligo

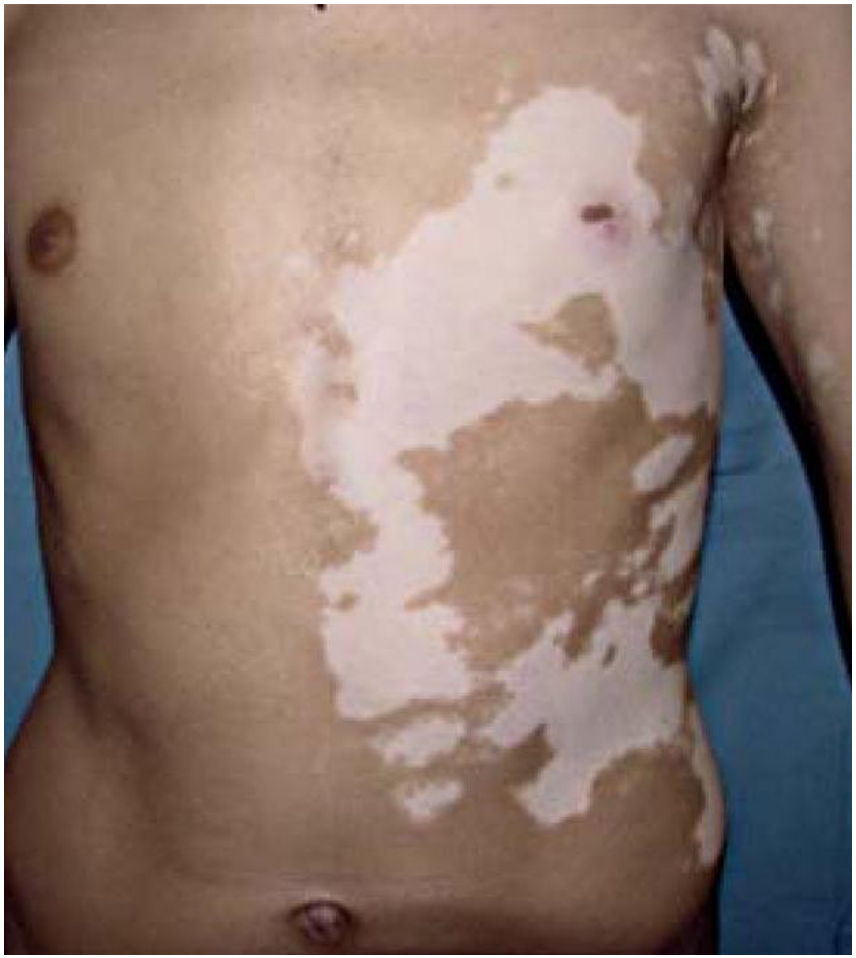
1. Focal
2. Unilateral (segmental)
3. Mucosal

Focal vitiligo



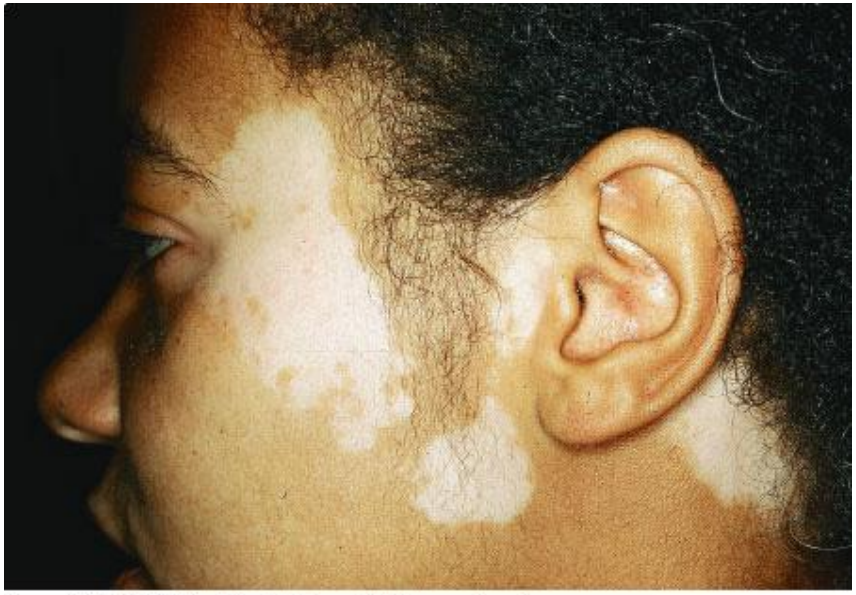
- one or more macules in one area
- not segmental distribution
- not evolved into generalized form after a period of 1-2 years

Unilateral (Segmental) vitiligo



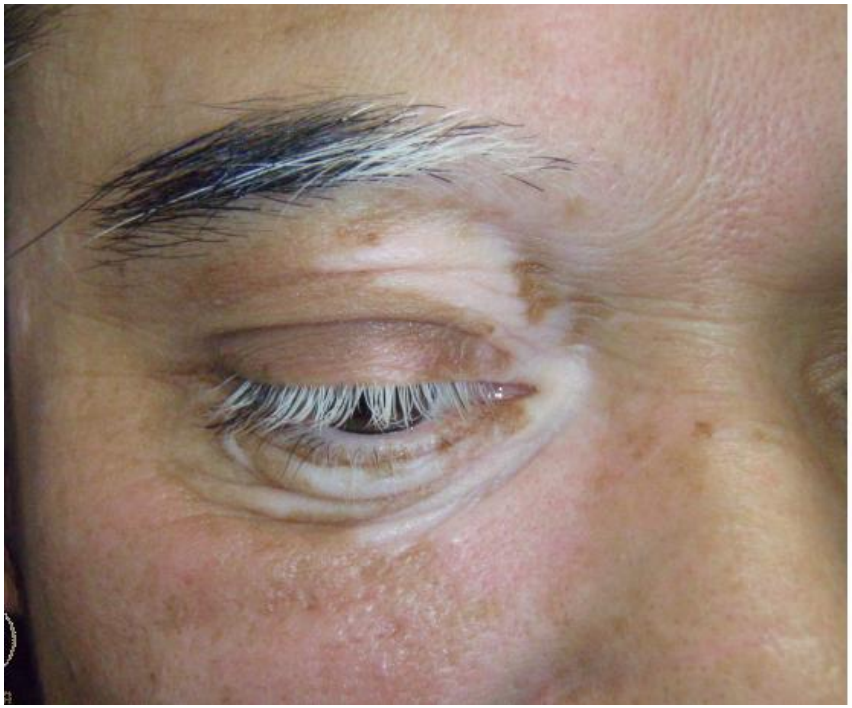
- an early onset in childhood
- one or more macules involving in an unilateral segment of the body, a dermatomal distribution or blashkoid pattern
- the macules does not cross the midline

Unilateral (Segmental) vitiligo



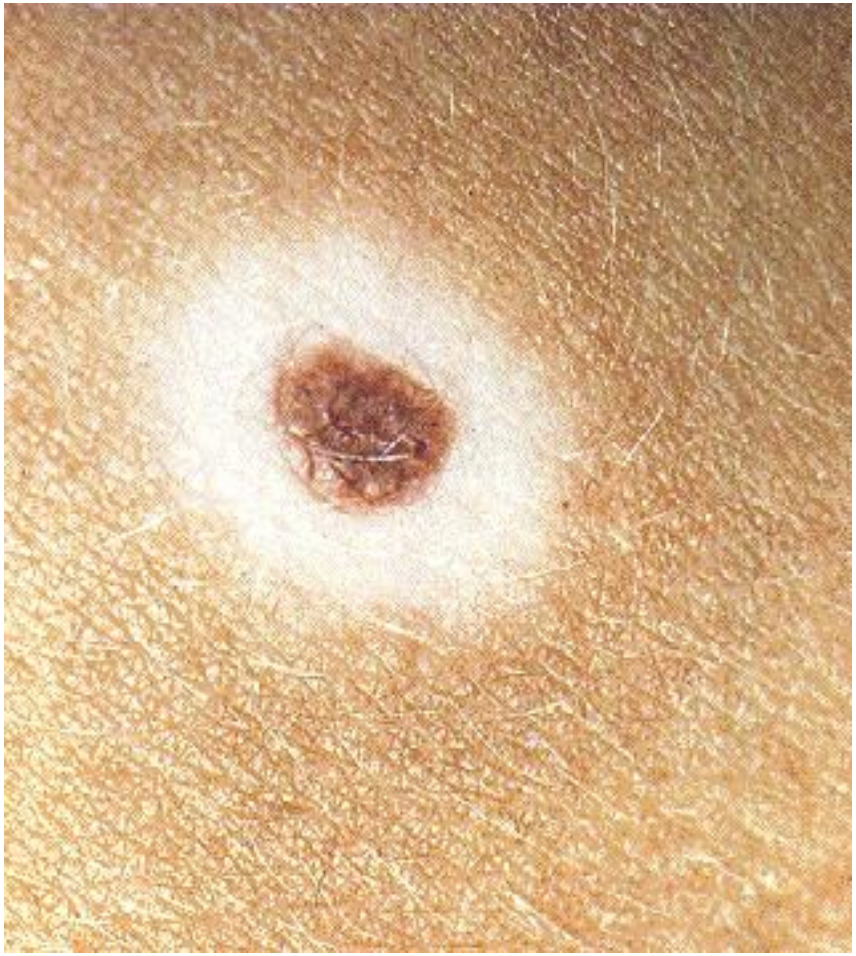
- Trigeminal area is the most common site of involvement
- The lesions develop rapidly over short span of time in a localized area and than remain stable

Unilateral (Segmental) vitiligo



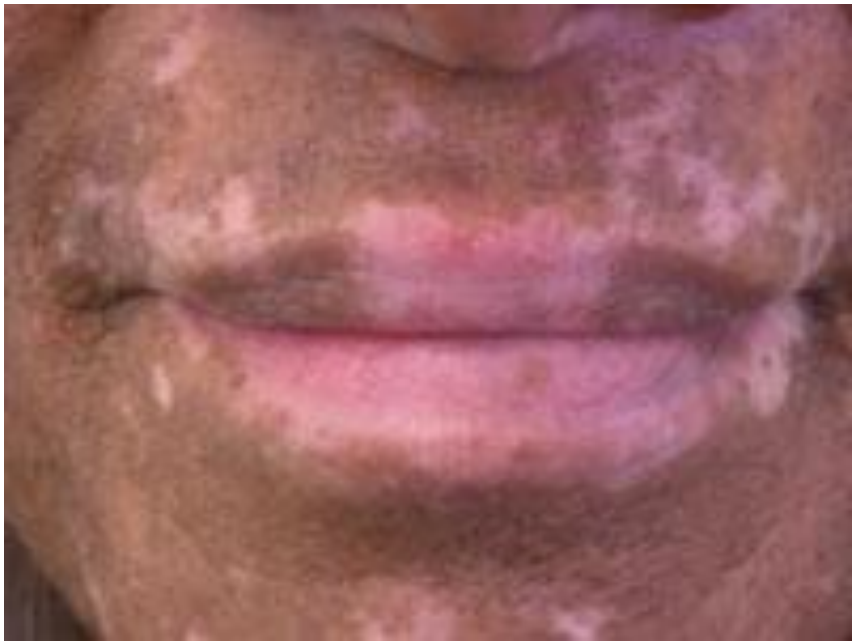
- Half of the cases are associated with poliosis
- Koebnerization not seen
- The lower incidence of family history of vitiligo and coexisting autoimmune disorders in SV has been reported.

Unilateral (Segmental) vitiligo



- The incidence of halo nevi is lower in SV
- Halo nevi is a marker for progression of SV to NSV

Mucosal Vitiligo

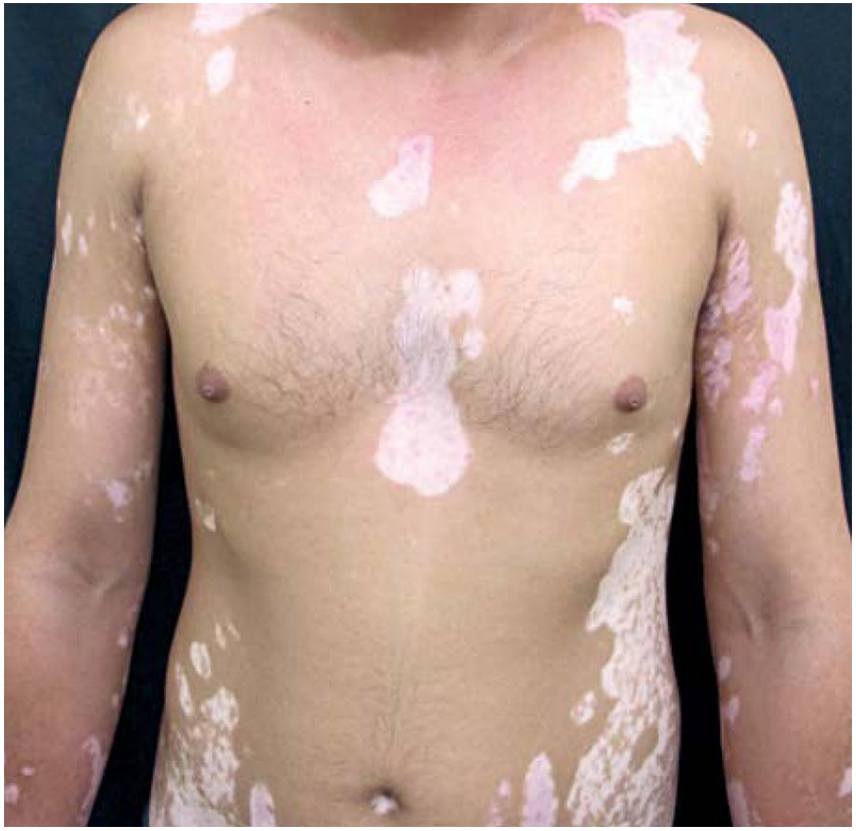


- lip involvement is the most common area
- involving any other mucosal structures (such as genitalia, nipples, gingiva)
- smoking is a precipitating factor
- mucosal involvement in generalized vitiligo is associated with progression of disease and also predicts poor prognosis

Generalized Vitiligo

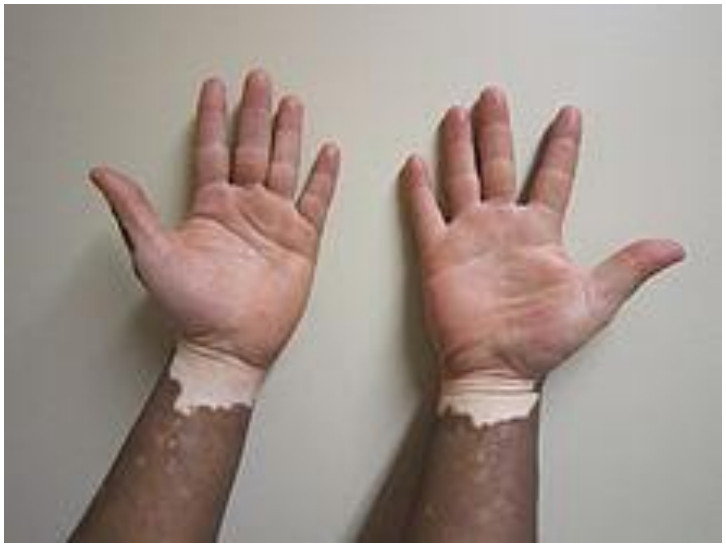
1. Vitiligo vulgaris
2. Acrofacial vitiligo
3. Mixed form

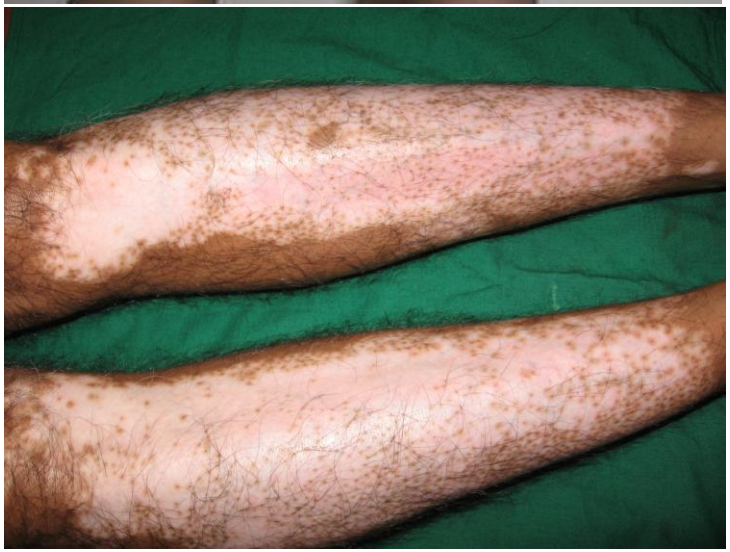
Vitiligo vulgaris



- The most common type of vitiligo.
- May begin later in life
- Characterized by few to many widespread macules
- Often symmetrically placed
- Seen at site sensitive to pressure, friction or trauma.
- The most common sites of involvement are fingers, wrists, axillae and groin, body orificies such as the mounth, eyes and genitals)

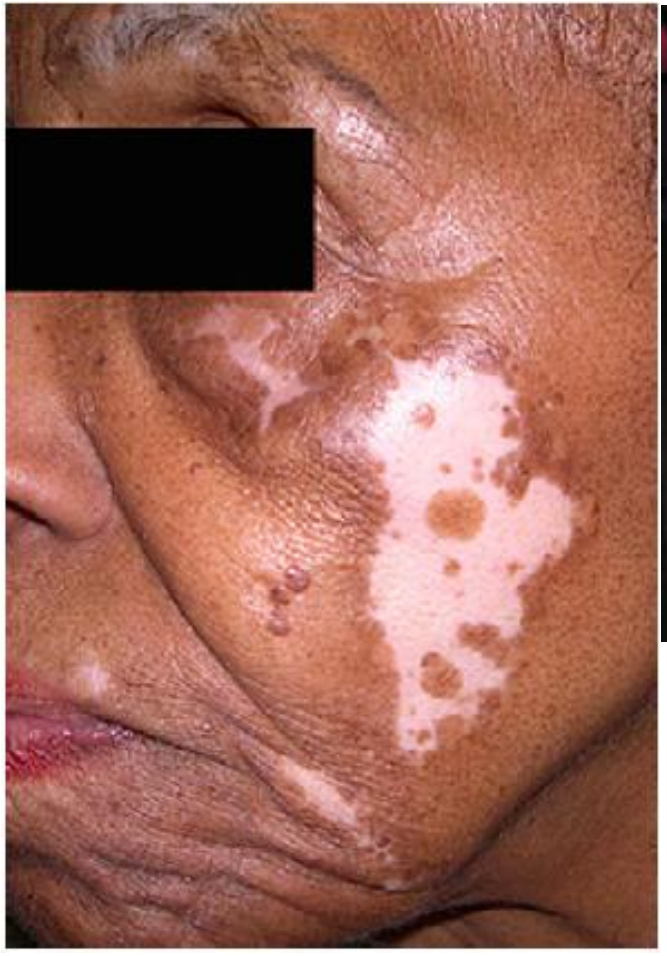
Vitiligo vulgaris

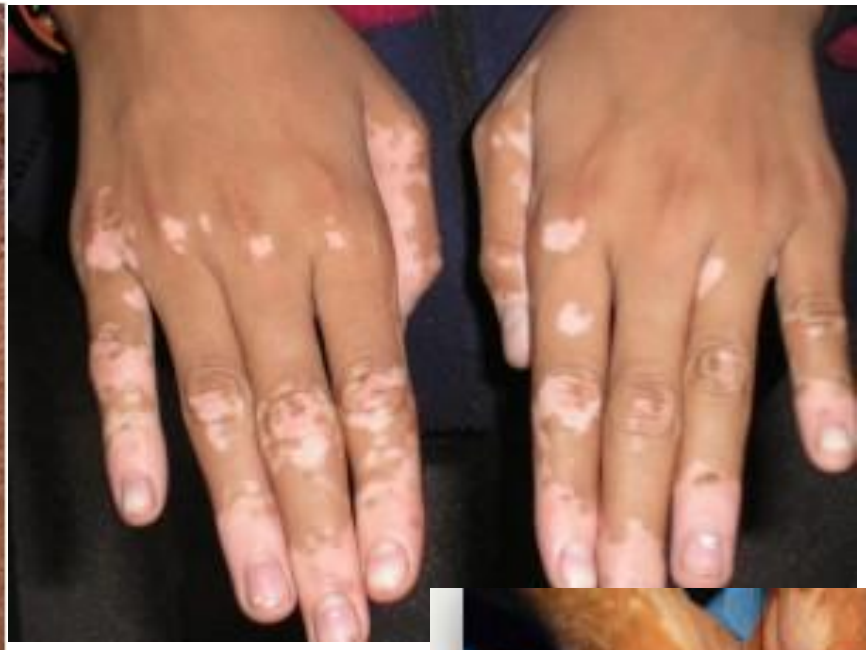


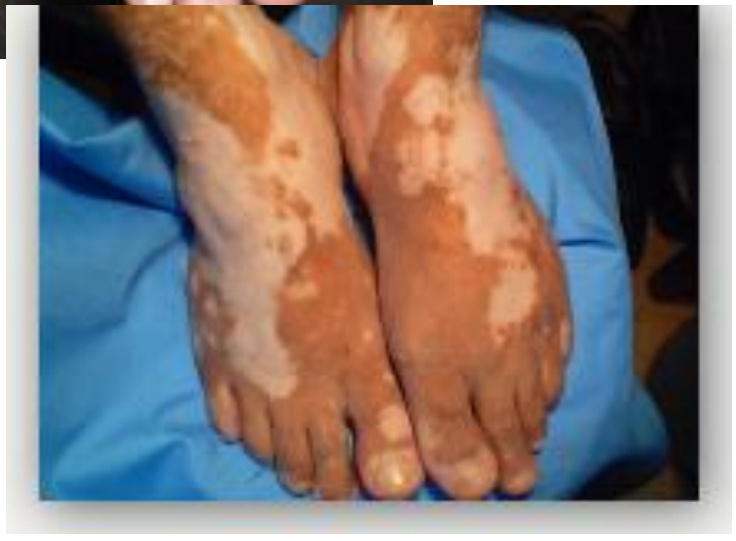


- typically progressive with flare-ups
- often associated with family history
- A higher incidence of autoimmune thyroid diseases
- Koebner phenomenon is commonly positive

Acrofacial vitiligo





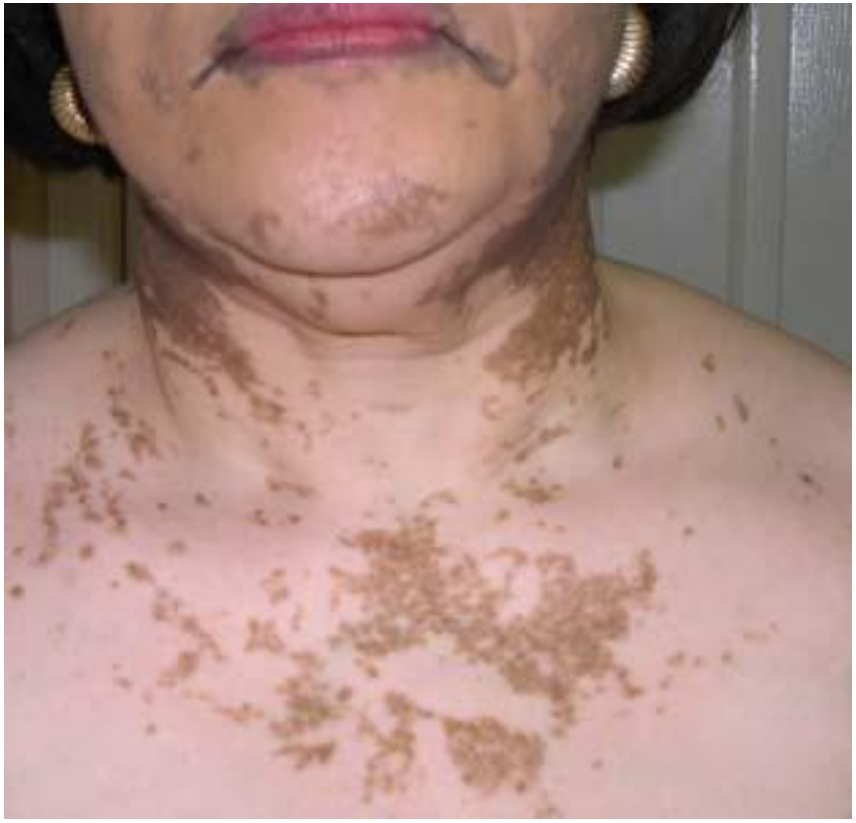


Mixed type

- More than one subtype of vitiligo in a patient

Universal Vitiligo

Universal vitiligo



- a few remaining normal macules
- Associated with the multiple endocrinopathy syndrome

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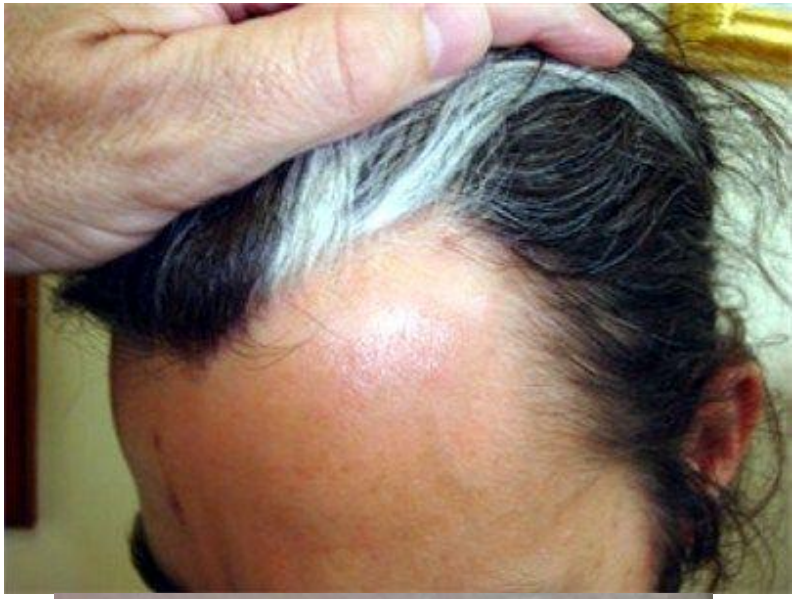
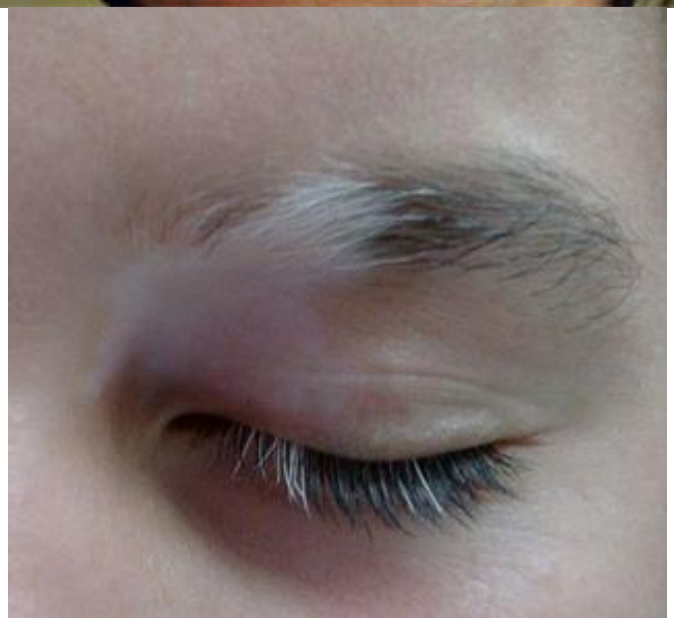
Cutaneous abnormalities in vitiligo

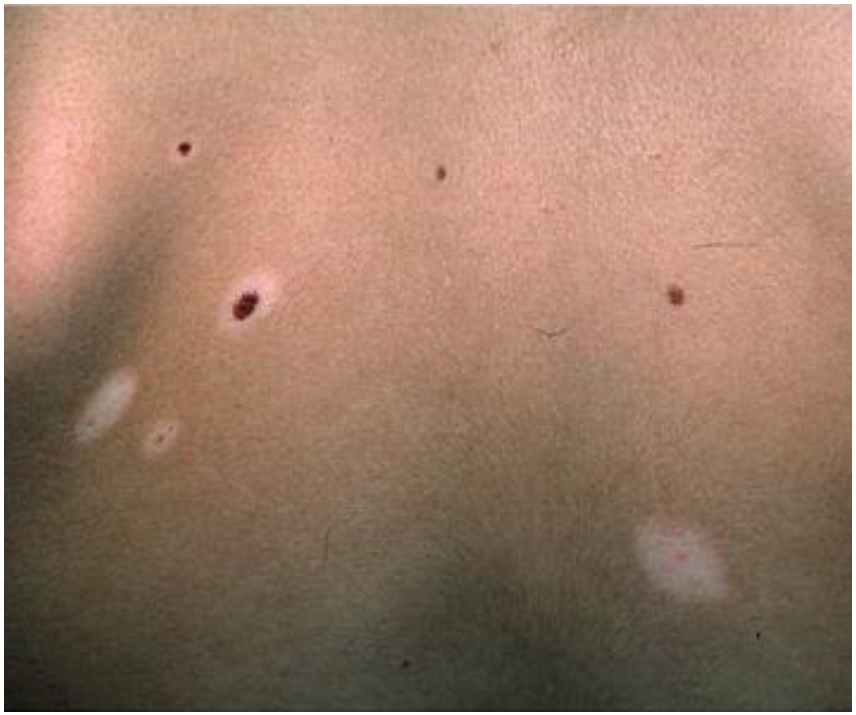
Leucotrichia (poliosis)





- Reported in 9 to 45 % of vitiligo patients
- Reported that extensive hair depigmentation may be a marker for poor prognosis

Halo nevi



- Commonly seen in vitiligo patients
- Confluence of these lesions in the stage of disappearance of the nevus leaves a typical vitiligo macule

Alopecia areata

- Reported in up to 16% of vitiligo patients

Vitiligo and ocular disease

- The most common ocular abnormality in vitiligo is uveitis
- The others are iris and retinal pigmentary abnormailities

Vitiligo and otic abnormalities

- Abnormal sensory hearing loss (suggesting an impairment of cochlear melanocytes)

Vitiligo and Systemic disease associations

More common associations	Less common associations
Addison disease Alopeci areata Atopic dermatitis **Autoimmune thyroid disease** Chronic urticaria **Diabetes mellitus** Halo nevi Hypoacusis Hypoparathyroidism Ichtiyosis Ocular abnormalities **Pernisious anemia** **Psoriasis** Rheumatoid artritis	Bazex syndrome Alezzandri syndrome APECED syndrome Asthma Ataxi-Telangiectasia Deafness Dysgamaglobulinemia HIV Inflammatory bowel disease Kabuki syndrome Kaposi sarcoma Melanoma MELAS syndrome Morphea Multiple sclerosis Myastenia gravis Nonmelanoma skin cancer Pemphigus vulgaris Sarcoidosis Scmidt syndrome Systemic lupus erythematosus Vogt-Koyagani-Harada syndrome

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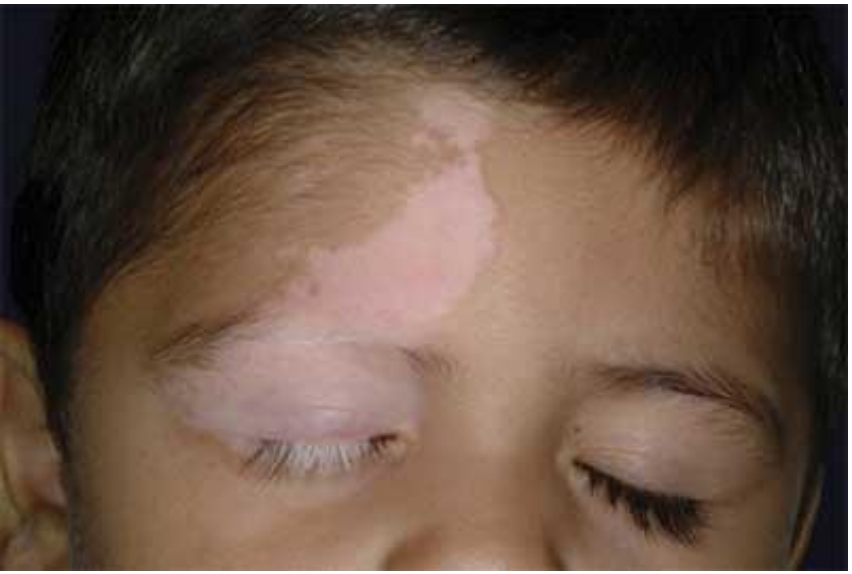
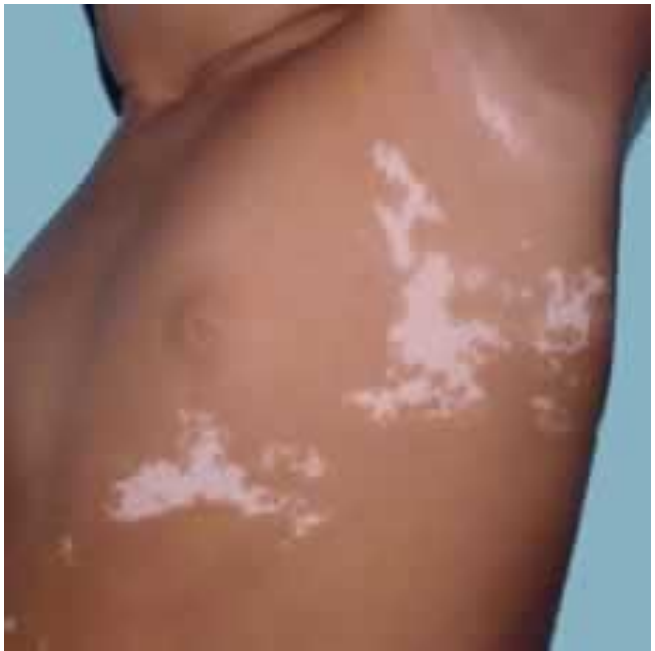
Childhood Vitiligo





- The frequency of SV is significantly increased in children compared with than in adults
- a slower rate of progression
- a higher prevalence of allergic diseases and lower prevalence of thyroid diseases
- children with vitiligo have been increased incidence of autoimmune antibody production
- more frequently in girls
- It has been reported that childhood-onset vitiligo had a significantly greater percentage of patients with a family history of vitiligo

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Course of vitiligo

- The natural course of vitiligo is unpredictable.
- The most common course is one of gradual evaluation of exisiting macules and periodic development of new lesions
- The onset of the disease is usually asymptomatic with appearance of a single or less often multiple macules.
- Following by progression for a time or a period of stability can be seen.
- Focal vitiligo may be a precursor of generalized vitiligo.
- Total spontaneous regression is rare.
- Segmental vitiligo is usually very stable.
- There is no consensus regarding the clinical evaluation of disease activity in vitiligo.

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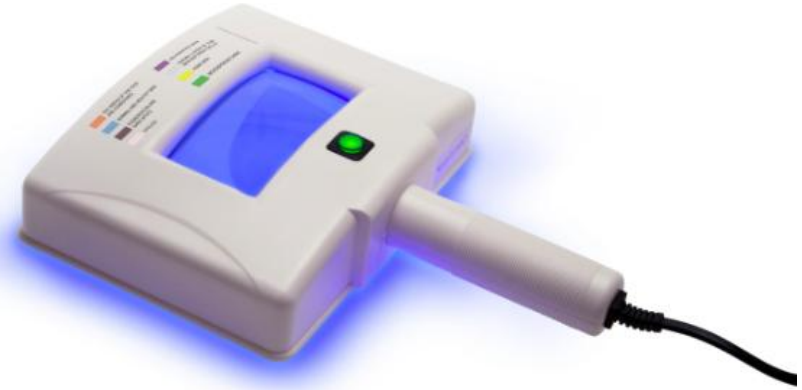
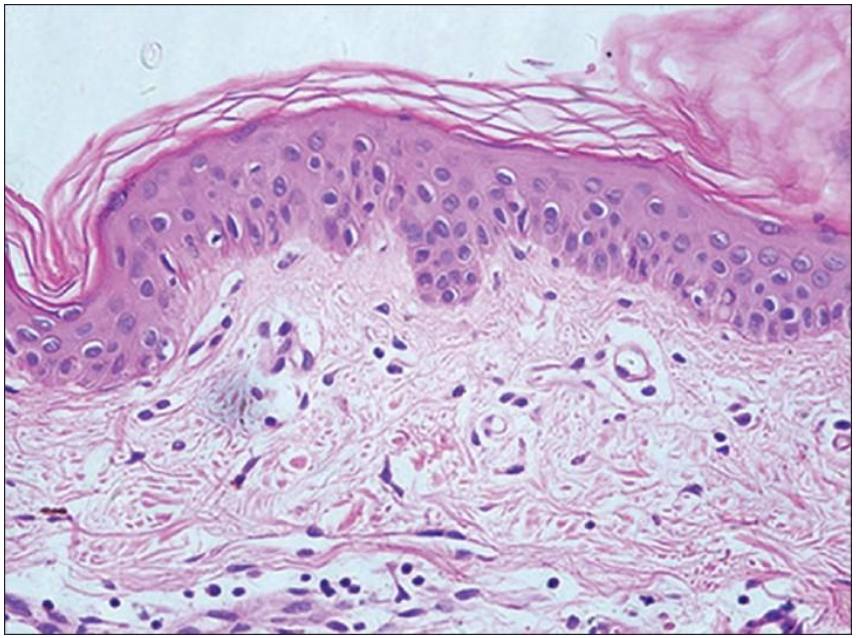
Diagnostic considerations



- The diagnosis of vitiligo is based on clinical examination
- Inspection with a wood lamp

Diagnostic considerations



- Biopsy is occasionally helpful for differentiating vitiligo from other hypopigmentary disorders

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Differential Diagnosis

Chemically-induced leokoderma (often occupational) Phenol and other derivatives	**Genetic syndromes** Hypomelanosis of Ito Piebaldism Tuberous sclerosis Vogt-Koyagani-Harada syndrome Waardenburg syndrome
Infections Leishmaniasis Leprosy Onchocerciasis **Tinea versicolor** Treponematoses	**Postinflammtory hypopigmentation** Atopic dermatitis Numuler dermatitis Phototherapy or radiotherapy indoced hypopigmentation Pityriasis alba Psoriasis Systemic lupus erythematosus Topical or systemic drug-induced hypopigmentation
Idiopathic **Idiopathic guttate hypomelanosis** Lichen sclerosus et athrophicus Progressive maculer hypomelanosis	**Malformations** **Nevus anemicus** Nevus depigmentosus
Neoplastic Amelanotic melanoma Mycosis fungoides	

Conclusion

Vitiligo;

- the most common depigmentation disorder,
- men and women are equally affected,
- half of the patients develop the disease before 20 years of age,
- classified into localized, generalized and universal,
- lesions typically develop in areas of friction (Koebner phenomenon),
- generalized vitiligo is the most common type,
- higher prevalence of thyroid diseases,
- association with ocular and otic abnormalities,
- the differential diagnosis is broad,
- the clinical course of disease is unpredictable

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THANK YOU