* Disorders of Pigmentations

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1- Normal skin and melanin definition and biosynthesis .

2- skin types .

3- Disorder of pigmentations .

4-Common Congenital hyperpigmentation

5-Acquired hyperpigmentation

6-Common Congenital hypopigmentation

7-Acquired hypopigmentation

*Our lecture talks about :

*Skin color factors :

* Hemoglobin (Pallor in anemia).
* Exogenous pigments in or on the skin surface, Carotinemia (orange in color) .
* Endogenously produced pigments (e.g. bilirubin).
*The pigments produced in the skin itself: melanin and phaeomelanin.

*Definition of Melanin

MELANIN an inert pigment which is produced by the **melanocytes** in the epidermal **basal layer**.

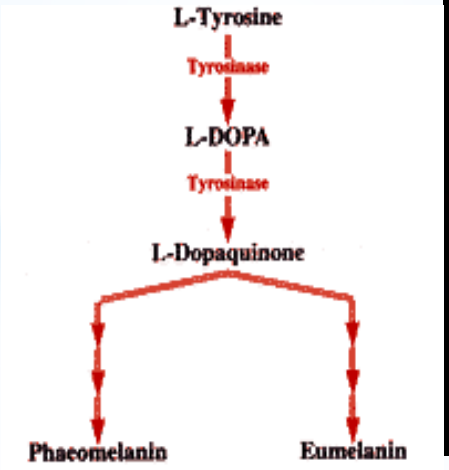
→Its function **to protect cell nuclei** from damage by UV which produce free radicals that damage the DNA and result in mutations that lead to carcinogenesis. defect in melanocytes increases risk of basal cell carcinoma

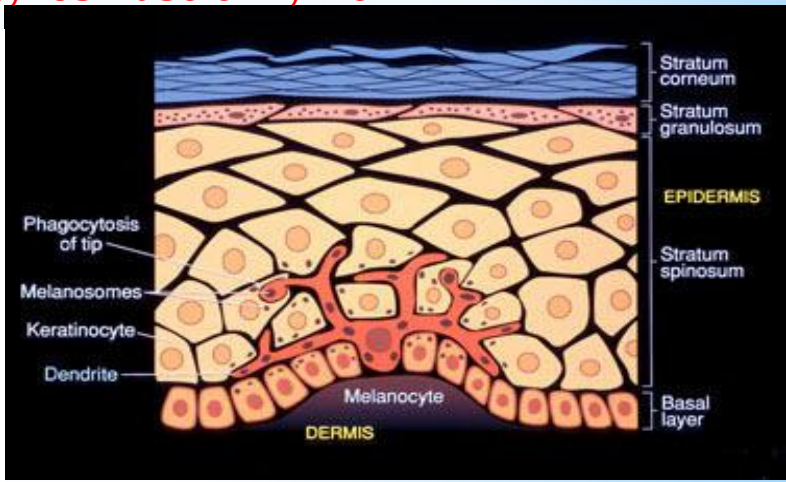
→It is synthesized from tyrosine in melanosomes by tyrosinase enzyme.

Eumelanin : deep brown –black

Pheomelanin: mainly in red hair

→ Melanocytes have dendritic distribution between stratum granulosum and spinosum , melanosomes inside these dendrites.





*Skin color

→The different skin colors result from the size and number of melanosomes but same number of melanocytes.

→Negro skin contains no more melanocyte than fair people.

In Jordan

*Disorders of pigmentations:

*Disorders of pigmentation can result from migration abnormalities of melanocytes from the neural crest to the skin during embryogenesis (Albinism).

*In addition, impairment of melanosome transfer to the surrounding keratinocytes.

*An alteration in melanin synthesis.

*A defective degradation or removal of melanin may lead to abnormal skin pigmentation.

*Immunologic or toxic mediated destructions of melanocytes can end in pigmentation disorders (Vitiligo).

→Disorders of pigmentation are classified in hypo- or hyperpigmentation which can occur as a genetic or acquired disease.

→They can manifest locally or diffuse.

*Common Congenital hyperpigmentation

1-Melanocytic nevi (Junctional , Dermal , Compound).

2-Giant congenital Melanocytic nevi.

3-Simple lentigines.

4-Café-au-lait macule.

5-Freckles.

6-The multiple lentigines syndrome.

7-Nevus of Ota.

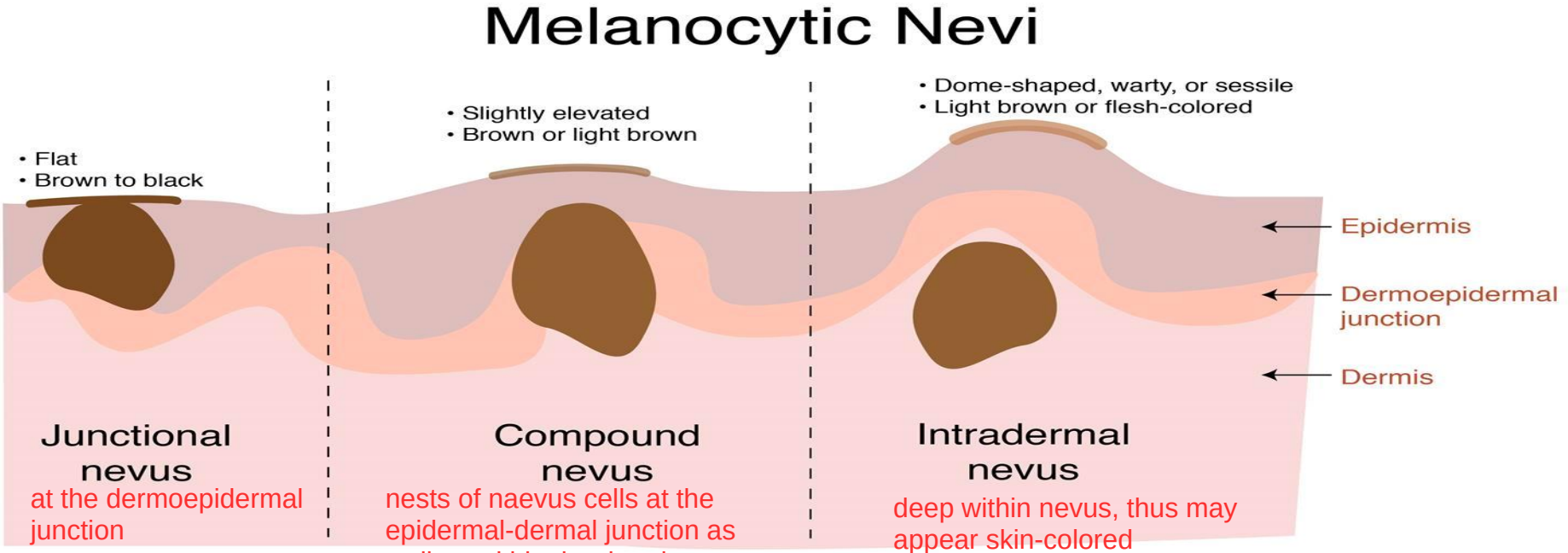
Brown , blue gray patch , usually inside , near or around the eye.

*Melanocytic nevi (moles)

→These spots may be flesh-colored to light-to-dark brown.

→They may be flat or raised.

Q!! Recount types of melanocytic nevi



*Melanocytic nevi (moles)

except?

→Although most moles are benign (non-cancerous) and will not cause any problems, some may change and become a skin cancer **called a melanoma especially the junctional type because they are more superficial.**

→For this reason, moles should be watched for bleeding, pain, itch, color, shape, symmetry, even borders, and size changes.

→No moles occur after 45 years of age , so after this age is considered melanoma until proven otherwise.

→Laser can treat the junctional type only.

ABCDE

- A way to check these moles is ABCDE:
  - **A**symmetry. If you divide your mole in half, both sides should look the same.
  - **B**order. The border of your mole should be even.
  - **C**olor. Your mole should be one color. Your mole should not have a variety of colors, especially colors like red or blue.
  - **D**iameter. Moles less than 0.6 cm in diameter are usually benign. If your mole increases in size, especially if it is greater than 0.6 cm, you should have it checked.
  - **E**volving or **E**levation. If your mole was flat but is now elevated (raised), or if you notice bleeding, crusting, pain, or itching, this should be checked out.

*Giant congenital nevi

*Congenital melanocytic naevi are usually classified by their size in an adult. There are several different classifications.

*A small congenital melanocytic naevus is < 1.5 cm in diameter.

*A medium congenital melanocytic naevi is 1.5–19.9 cm.

***A large or giant congenital melanocytic naevus is ≥ 20 cm in diameter , treated by Plastic surgery (FULL EXCISION).**

*Simple lentigines

***Simple lentigines** are the result of increased melanocytes in the stratum basale layer of the skin and sometimes increased melanin content in the upper layers of the epidermis and stratum corneum , **brown in color**.

**They can occur anywhere on skin or mucous

*Café-au-lait spots

a solitary cafe-au-lait spot is not suspicious for malignancy

***Six or more spots of at least:**

***5mm in diameter in pre-pubertal children.**

***15mm in post-pubertal individuals is one of the major diagnostic criteria for diagnosing Neurofibromatosis.**

→ In 95% of NF1 , these spots are the first sign .

*Freckles النمش

→Freckles are small , harmless marks that appear on the skin. Genetics and sun exposure are the primary causes of freckles.

*Some people are more likely to get freckles than others, depending on their genes and skin type.

*Freckles are common in children and may disappear or become less noticeable as they grow up.

*People with dark hair, eyes, and skin usually produce mostly eumelanin and are less likely to develop freckles.

*People with red, blonde, or light brown hair and who have light-colored skin and eyes usually produce mainly Pheomelanin and are more likely to develop freckles.

*People can prevent or reduce the appearance of freckles by protecting their skin from the sun.

*Protecting the skin from sunlight will not reduce the appearance of existing freckles, but it can prevent new freckles from forming.

Treated by laser ?
NO

*Acquired hyperpigmentation

Addison ’ s disease.

Renal failure.

Haemochromatosis.

Liver disease.

Carotinemia:

– Idiopathic.

– Myxoedema.

– pernicious anemia.

– Antiepileptic drugs , Food (carrot).

Acanthosis nigricans.

Chloasma.

Drugs and chemicals.

Post inflammatory hyperpigmentation.

*Acanthosis Nigricans

***Acanthosis nigricans** is a medical sign characterized by brown-to-black, poorly defined, velvety hyperpigmentation of the skin. It is usually found in body folds, such as the posterior and lateral folds of the neck, the armpits, groin, navel, forehead and other areas.

***Def : Thick velvety skin in a skin fold.**

→ Thin person with AN , think of gastric adenocarcinoma .

*Melasma (Chloasma)

*Melasma (also known as Chloasma) is marked by **tan or brown patches** that may appear on the **forehead, cheeks, upper lip, nose, and chin**.

*Although this condition is often called the **"pregnancy mask,"** men can also develop it (genetics , type III , IV).

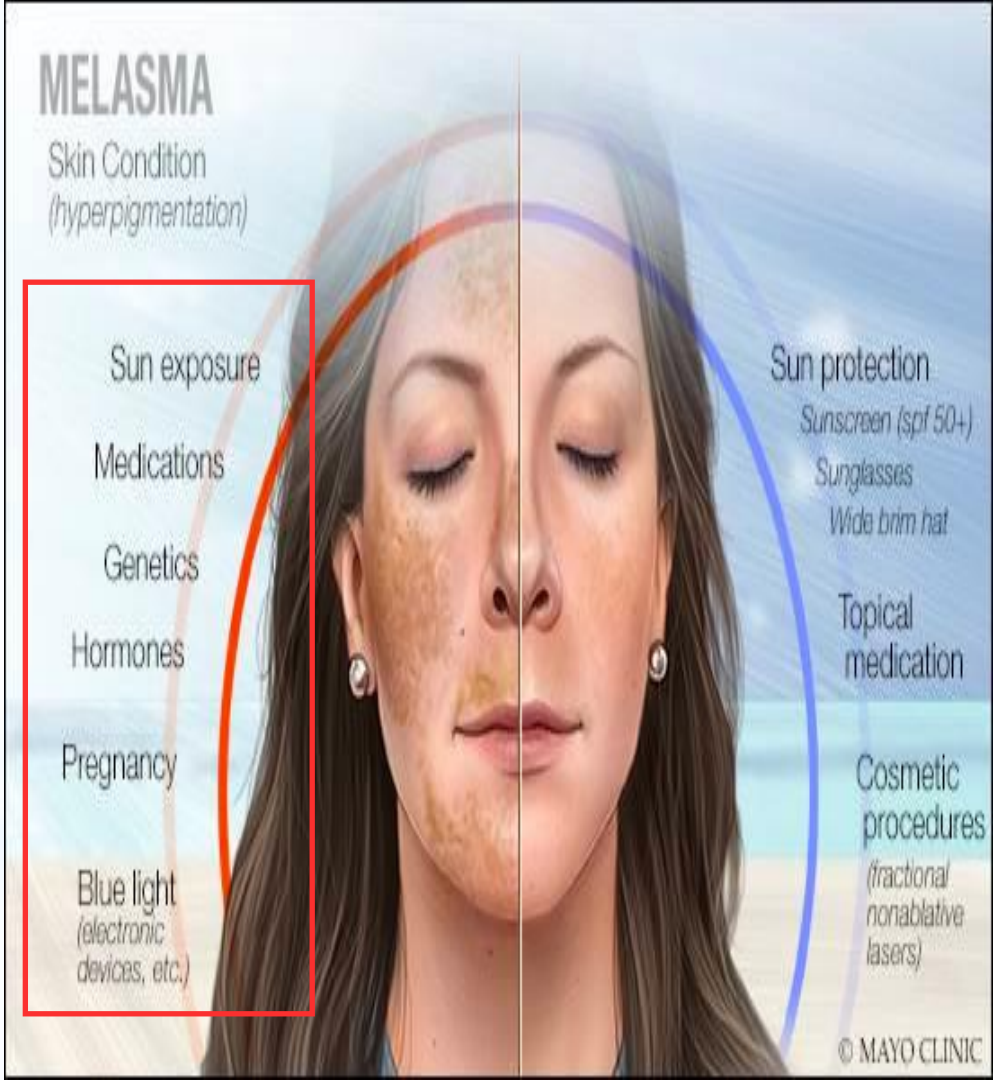
***It may also occur in women who are taking birth control pills or postmenopausal estrogen.**

***Melasma may go away after pregnancy, but if it remains, it can be treated with certain prescription creams and some over-the-counter skin care products.**

***In addition, lasers that target pigment can be helpful.**

***Sunscreen use at all times because sunlight will make the condition worse.**

*** Diagnosed with wood's lamp .**



*Post inflammatory hyperpigmentation

Q!! Recount causes of post inflammatory hyperpigmentation

Lichen planus.

Fixed drug eruption.

Eczema.

Acne

Repeated exposure of drug ,→ new lesion → light up the old one .
Usually after one week .
Antibiotics , anti-epileptics .
Usually after one week .
Mostly mucosal patch

*Hypopigmentation

Congenital:

- Albinism.
- Phenylketonuria.
- * Piebaldism
- Tuberous sclerosis complex.
- Hypochromic naevi (no depigmentation so its not vitiligo).

Acquired:

- Vitiligo.
- Sutton ’ s halo naevi..
- Tuberculoid leprosy.
- Pityriasis (tinea) versicolor.
- Pityriasis alba.
- Lichen sclerosis.
- Drugs and chemicals.
- Post inflammatory hypopigmentation.

*1- congenital :Hypomelanosis (generalized)

Albinism (البرص)

***Defect of melanin production results in little or no color in skin ,hair and eyes**

***Due to congenital inability to form melanin patients have fair skin blonde hair and pink irises.**

***Have poor vision photophobia.**

***Increase risk of developing skin Cancer.**

1-Oculocutaneous albinism:

Severe form.

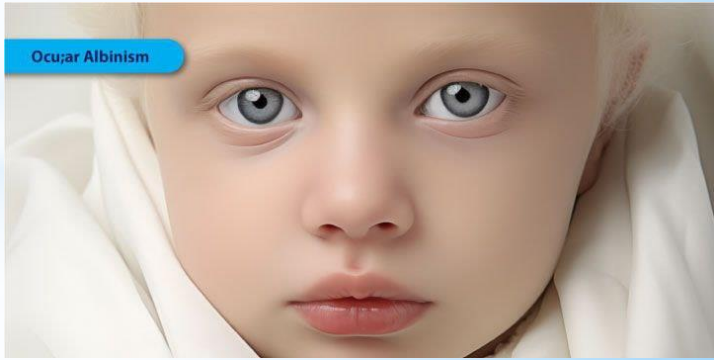
Have white or pink hair, skin and iris colo.r

Vision problem.

2-Ocular albinism:

Rare,Only the eyes.,No coloring in retina.

Skin and eye colors are normal.



*Piebaldism

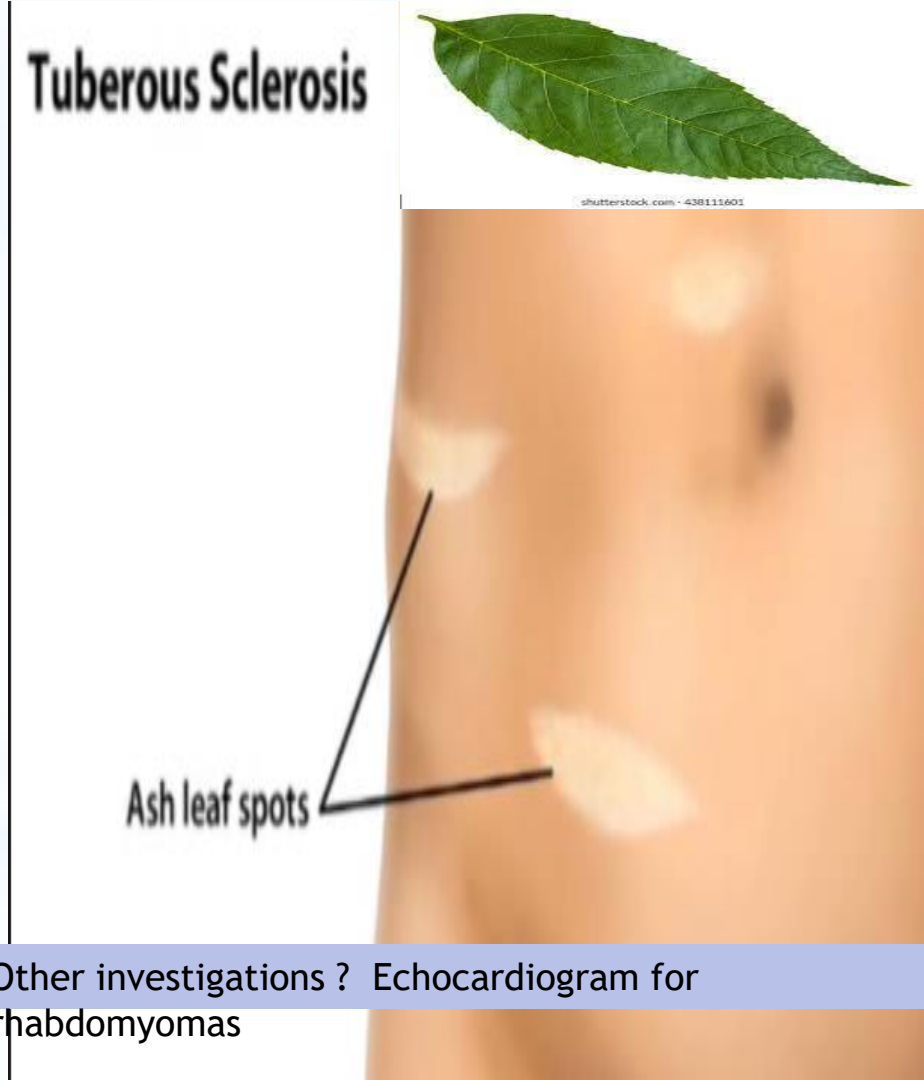
- **Piebaldism is a genetic condition, typically present at birth, in which a person develops an unpigmented or white patch of skin or hair.**

*Systemic Associations:

*Localized hypo- or hyperpigmentation in children may serve as markers for systemic diseases.

***Ash-leaf hypopigmentation are characteristic for tuberous sclerosis.**

***<u>The most common autoimmune-induced depigmentation is vitiligo.</u>**



Other investigations ? Echocardiogram for rhabdomyomas

*Achromic naevus (hypochromic nevus)

*Achromic naevus **(nevus anemicus)** is an uncommon birthmark characterized by a well-defined pale patch.

*This is usually several centimeters in diameter, with an irregular but well-defined border.

Shape and size varies. Often, smaller **hypopigmented macules** arise around the edges, resembling a splash of paint.

Negative in wood's light.

Acquired

* Vitiligo (البهاق)

***Skin condition resulting from loss of pigment which produces white patches.**

*It may be an **autoimmune process.**

*The peak age of incidences the **2nd and 3nd** decade.

*Any part can affected usually **both sides of body.**

Common area :face, lips ,hands, arms ,legs.

*Typical vitiligo shows area of **milky-white skin well demarcated patch , we can use wood's lamp .**

Vitiligo often begins with a rapid loss of pigment ,this may continue until for unknown reasons the process stops.

*Most people are in good health although may occur with other autoimmune **disease as thyroid and other autoimmune diseases.**

So investigations include : anti-TTG , anti-TPO , FBS , ANA…etc.

should always rule out any autoimmune disease.

*Sometime the best treatment for vitiligo **is no treatment** at all, Because these area are easily sun burned.

but it DOES have treatments!!!

*They have a risk to **skin cancer** they should wear a sunscreen, avoid sun exposure

***Treatment can be aimed at returning normal pigment(repigmentation)or destroying remaining (depigmentation)**

→**Repigmentation therapy** :topical corticosteroid for small areas of vitiligo these agents can thin the skin or even cause stretch marks in certain areas

None of the Repigmentation therapy are permanent cures☹ ☹ ☹.

PUVA is a form of regimentation therapy where the type of medication known as **psoralen** which makes the skin very sensitive to light then the skin is treated with special type of ultraviolet light called UVA

Efficacy between 50-70% on face trunk and upper legs ,upper arms but hands and feet response poorly

*Usually at least a year of twice weekly treatment are required

*Long term used leads to :freckling of skin.

*Increase risk of skin cancer.

C.I:

Children less 12

Pregnant

Breast feeding

→**Narrow band UVB**, This is a form of phototherapy that requires the skin to treated 2-3 times a week for a few months. At this time it's not widely available. Useful in treating children

*Depigmenations

*For some patients with extensive involvement

easier to remove smaller brown areas than repigment the larger areas of vitiligo
think: Michael Jackson

*The most practical treatment is to remove remaining pigment from normal skin and make whole body an even white color

*Done with a chemical called; **Monobenzylether** take a one year to complete

*The pigment removal is permanent

* Sutton ' s halo naevi

*A halo naevus is an otherwise normal mole with a white ring, or halo, around it.

*The central dark brown naevus fades from dark brown to light brown to pink, eventually disappearing completely , ***needs follow up for melanoma.**

***Halos can be seen as part of a more generalised pigment loss, vitil igo elsewhere, and halo naevi may also be associated with another autoimmune disease.**

*Post inflammatory hypopigmentation

Q!! Recount types of post inflammatory hypopigmentation:

*Tinea versicolor (M.furfura destroys lipids releasing fatty acid (Azeliac acid) leading to irritation of melanocytes).

*Pityriasis alba , can't be seen in wood's lamp .

*Psoriasis (Short Melanocyte transit time, also topical steroids can lead to hypopigmentation).

Pityriasis versicolor

Psoriasis

➢ **Pityriasis alba**

✓ **Ill-defined hypopigmented patch , more in dark skin and atopic dermatitis.**

✓ **Ttt : Emollient + Avoid sun exposure , steroid if inflammatory**