THE PROPOSED CLASSIFICATION OF HYPO AND DEPIGMENTARY SKIN DISORDERS

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Hypo or depigmentations observed clinically can be due to the defect in melanogenesis or by the faulty transfer of melanin to epidermal cells. Rapid dropping off, of the pigment into the corium, may be another contributory factor[1] (Graham et al.). Relative lack or complete absence of pigment is quite commonly encountered in our country and due to contrast in colour it constitutes a major cosmetic problem. In India unfortunately since ages the taboo of leprosy is associated with leucoderma (Lahiri). In some parts, vitiligo or leucoderma is considered as white leprosy.

Classification, no doubt, facilitates the basic understanding of the subject but it is usually a controversial problem. We are herewith presenting our classification on the etiological basis of various hypo and depigementary skin disorders.

In our opinion, hypo and depigmentary disorders of the skin should be classified into ten groups as given below:

1. Congenital
2. Idiopathic
3. Chemical
4. Physical
5. Endocrinal
6. Nutritional
7. Metabolic
8. Achromias due to infections
9. Post-inflammatory
10. Miscellaneous and iatrogenic

1. *Congenital*: In this group we include.
   1. Complete albinism,
   2. Incomplete albinism
   3. Albinoidism (leucism) and
   4. Local Albinism (Nevus Achromicus)

Albinism[2] denotes congenital absence of pigment in the skin, hair and choroid.

1. Complete albinism, with its lack of pigment and striking ocular changes, has a recessive hereditary pattern.

2. Incomplete albinism has some pigment in the skin and hair and presents ocular manifestation as in complete albinism. Nystagmus is usually present and this type may be determined by a recessive or irregularly dominant gene.

3. Albinoidism ' or leucism is another variant of albinism and is controlled by a dominant autosomal gene. Albinoids have normal eyes.

4. Local albinism is transmitted as a dominant Mendelian characteristic with complete penetrance (Butterworth). This condition is characterized by localized

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depigmented areas present at birth and remaining constant throughout the life. In some cases, depigmentation has been noted in distribution comparable to that of a linear nevus or nevus unius lateris[3] (Sulzberger). Partial albinism, congenital vitiligo, congenital achromia, nevus achromicus[4] (Behl) piebaldness and white nevus[5] (Lahiri) are the various synonyms used for this condition. In Klein[6] and Waardenburg Syndrome, partial albinism is associated with deaf-mutism, hypoplasia and heterochromidia of the iris and certain other anomalies. Local albinism should not be mistaken for nevus anaemicus characterized by the congenital absence of normal blood supply to a patch of skin appearing as a light coloured area[7] (Pillsbury). In these lesions the amount of melanin is normal[8] (Andrews) while the triple response of Lewis is absent.

2. The Idiopathic group comprises of
   1. Vitiligo
   2. Vogt-Koyanagi Syndrome
   3. Perinevoid vitiligo or Halo nevus
   4. Dyschromatosis symmetrica hereditaria
   & 5. Pityriasis Alba or Pityriasis simplex Facei

1. Vitiligo or achromia (Ormsby) is an idiopathic, *hereditary* acquired progressive loss of pigment due to the failure of function of the tyrosinase system of the melanocytes (Butterworth). The exact cause of this functional disturbance is not known (Lewis). This defect appears to be transmitted as an autosomal dominant Mendelian characteristic (Butterworth). Vitiligo might be unilateral and segmental.

2. Vogt-Koyanagi Syndrome : In this syndrome, first suggested by Babel[9] in 1939, vitiligo is found in association with bilateral uveitis, dysacousia, premature greying of hair and alopecia areata (Lever). Achromic lesions are found in about half the cases of this syndrome (Andrews). Although virus and allergic theories of its origin have been propounded, the exact cause as yet remains undecided[10] (W. C. Johnson). In the past decade, the term Vogt-Koyanagi-Harada syndrome has been used to describe this idiopathic condition[10].

3. Perinevoid vitiligo or Halo nevus : Sutton, in the year 1916, first described this idiopathic condition as leukoderma acquisitum centrifugum. In perinevoid vitiligo (Andrews) one observes depigmentation around one or more pigmented nevi (Butterworth). The most recently held view is that this idiopathic entity is no more than a variant of vitiligo[12] (Frank S. B. et al).

4. Dyschromatosis Symmetrica Hereditaria : It is an idiopathic anomaly of pigmentation occurring among the Japanese. It is characterized by spotted pigmentation mingled with depigmentation at the ends of extremities and is transmitted as an autosomal dominant trait (Butterworth,).

5. Pityriasis Alba or pityriasis simplex facei is a commonly encountered idiopathic pigmentary disorder. It is characterized by the formation of round or oval hypochromic macules with fine scales mostly appearing on face, neck, shoulders and upper arms of children. Although according to Sulzberger et al, it is probably due to

superficial streptococccal infection, no evidence to support the role of streptococci, other bacteria or fungi has been yet obtained[13] (Bassaly et al. & Wells et[14] al.). Parasitic infestations and vitamin deficiencies were suggested as causative factors by Tas J.[15] Other synonyms used for this idiopathic condition are impetigo furfuracea and dartres volantes.

3. *Synthetic & Chemical Leucodermas* comprise of secondary hypo and depig–mentations produced due to.

1. Monobenzyl ether of hydroquinone (M. B. H.)
2. Hydroquinones & quinoid substances
3. Photographic developers
4. Phenol-formaldehyde resins
5. Guanofuracin
6. Nylon (Hexamethylene diamine condensate of adipic acid)
& 7. Superficial burns due to chemicals.

Oliver[16] and his associates, in the year 1939, first reported a peculiar occupa–tional leukoderma occurring on the hands of tannery workers using heavy rubber gloves. It was proved to be due to antioxidant monobenzyl ether of hydroquinone incorporated in the rubber to improve its durability. Depigmentations have also been reported due to contact with rubber foot wear[5] rubber garters (Andrews) and rubber condoms (B. Solomon & Andrews). The exact mechanism by which monobenzyl ether of hydroquinone affects melanogenesis is obscure[18] (Snell R. S.). Denton et al,[19] suggested that the substance is either converted into hydroquinone which in turn inhibits melanogenesis by inhibiting the conversion of tyrosine to dopa or prevented the oxidation of SH groups which bind copper in the tyrosinase complex. Symmetrical vitiligoid lesions have been reported by Chumakov[20] et al. in workers of phenol–formaldehyde resin industry in Russia.

Depigmentations have also been produced by other quinones, quinoid substances, (Andrews) photograpic[21] developers (Clarke G. H. V.) and nylon watch straps[22] (M. El Zawahry). Paul et al· (Andrews) and Prof. Kitamura[23] have reported leuko–derma on eyelids caused by the topical use guanofuracin containing eye lotion. Superficial chemical burns due to loss of melanocytes often lead to hypo or depigmentations. Due to day by day increasing contacts with various chemicals and synthetics, the incidence of this groups of achromias and hypochromias is likely to increase remarkably in near futnre.

4. *Physical Leukodermas* include hypo and depigmetations due to
1. Superficial thermal burns.
2. Pressure caused by tight wearing of garments e. g. dhotis or sarees.
3. Superficial abrasions due to external trauma.
4. Cold.

In all these conditions, the local damage to melanocytes is the causative factor. Leucoderma around waist due to the tight wearing of the sari is quite commonly enco–

untered in Indian women. The cold climate also retards melamogenesis and produces diffuse hypopigmetation.

5. *Endocrinal achromias* include hypopituitarism[24] and generalized myxedema[25] (Sulzberger) leading to diffuse hypopigmentation and Addison's disease of vitiligoid type leading to patchy depigmentation. Sir D. Dunlop[26] has reported pigmentation in Addison's disease alternating with leucodermic patterns.

6. *Nutritional Achromias* include Kwashiorkor's Syndrome and depigmented or hypopigmented lesions due to para-amino benzoic acid deficiency (B. Solomon). In Kwashiorkor's Syndrome caused by lack of vitamins and proteins, besides crazy pavement dermatosis and oedema, depigmentations[27], [28] and hyperpigmentations of the skin may occur.

7. *Metabolic Achromia* includes vitiligoid type of primary cutaneous amyloidosis. Jolio. M. Borda[29] has reported two cases of this type of histologically proved primary cutaneous amyloidosis with lesions situated on the anterior aspect of the legs and having hypochromia as a presenting feature.

Metabolic achromia also includes sclero-vitiliginous type of porphyria reported by Rogailin[30] in 1962. Of 150 patients suffering from porphyria observed by the author for several years, 5 developed typical sclero-vitiliginous lesions.

8. Hypochromias and achromias occur in below mentioned bacterial, treponemal and protozoal infections :-

1.	Bacterial :	Leprosy
2.	Treponemal :	Syphilitic leucoderma (Leukoderma Colli)
		Yaws
		Bejel
		Pinta & Hemi-pinta
3.	Protozoal :	Post Kala-azar Dermal Leishmaniasis

In leprosy hypochromic lesions are found in maculoanaesthetic, indeterminate and lepromatous forms. Typical well-defined hypochromic and anaesthetic macules are however found only in the maculo-anaesthetic variety.

Leukoderma Colli or collar of venus constitutes depigmented areas resembling milk spots on neck of women coincident with the appearance of the macular eruption of secondary syphilis or within several months following it (Andrews).

Vitiligo like depigmented areas formed in the tertiary stage of yaws which is endemic, non-venereal treponematosis caused by Treponema pertenue (Willcox). In India, Yaws is endemic in the Bastar district of Madhya Pradesh.

Depigmented lesions also appear in the late stage of Bejel (Willcox) which is probably a variety of nonvenereal syphilis seen in Bedouin Arabs (Ambrose King).

Pinta is non-venereal tremonemal disease caused by T. carateum and characterized by bizarre pigmentary and depigmentary changes. Persistent symmetrical vitiligo-like lesions appear on the body. White pinta represents the terminal stage of depigmenta-

tion (Andrews). Hemi-pinta[31] is a rare variety in which pigmentary disturbances affect only the half side of the body (Andrews).

Post-kala-azar Dermal Leishmaniasis[32] was first reported in the medical literature in 1922 by Brahmachari in Calcutta. It occurs in areas where Kala-azar is endemic and may lead to the formation of depigmented spots or erythematous patches of various sizes, and yellowish pink papules or nodules. Skin snip for L. D. bodies is usually positive from nodular lesions and is usually negative if taken from depigmented lesions.

9. *Post-inflammatory Leucodermas*: After involution of some inflammatory skin disorders especially infections, due to either toxic inhibition of melanogenesis or due to the interception of the ultraviolet rays by the skin lesion acting as a screen hypo or depigmented lesions are formed (Lewis) either at site of the original lesion or around it as a depigmented halo (Fitzpatrick).

Consecutive or post-inflammatoay leucodermas may follow: Tinea versicolor or pityriasis versicolor
Psoriasis
Seborrhoeic dermatitis
Para-psoriasis (Tobias)
Neuro Dermatitis Circumscripta (Sulzberger)
Atopic dermatitis
Pityriasis rosea
Impetigo contagiosa
Herpes Zoster (Pillsbury and Andrews)
Radiodermatitis
Arsenical or gold dermatitis (Sulzberger)

10. *Iatrogenic & Miscellaneous*: Pillsbury[33] has mentioned depigmentation occurring in patients receiving BAL (2,3-dimercaptopropanol) and Thiouracil while according to Sulzberger[34] quinacrine hydrochloride (Atabrine) may lead to hyper or depigmentation.

Some of the other disorders in which hypo or depigmentation is likely to occur are:

1. Scleroderma (Circumscribed as well as systemic)
2. Chronic discoid L. E.
3. Sclero-cicatrical type of lichen planus of scalp (J. M. Borda)[35]
4. Epidermolysis bullosa dystrophica
5. Lichen sclerosus et atrophicus (Lichen albus von Zumbush)
6. Xeroderma pigmentosum end stage (Lewis)
7. Anetoderma (Behl)
8. Pseudo-atrophoderma colli (B. Solomons)
9. Poikiloderma of Civatte (B. Solomons)
10. Pseudopelade of Brog

As in the literature we could not find any conclusive evidence of hypochromic or achromic lesions produced directly due to allergy, we have not included this group in our classification.

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TABLE SHOWING THE CLASSIFICATION OF HYPO & DEPIGMENTARY SKIN DISORDERS

Proposed by
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Congenital	Complete albinism, incomplete albinism, albinoidism and nevus achromicus.
Idiopathic	Vitiligo, Vogt. Koyanagi Syndrome, Halo Nevus, Dyschromatosis symmetrica hereditaria and pityriasis alba.
Chemical & Synthetic	Monobenzyl ether of hydroquinones and quinoid substances, photographic developers, phenal formaldehyde resins, guanofuracin and Nylon.
Physical	Superficial thermal burns, pressure due to tight wearing of garments, superficial abrasions and cold.
Endocrinal	Hypopituitarism, Generalized myxoedema, Addison disease vitiligoid type.
Nutritional	Kwashiorkor's Syndrome.
Metabolic	Vitiligoid Type of primary cutaneous amyloidosis and sclerovitiliginous type of porphyria.
Bacterial	Leprosy.
Treponemal	Leucoderma colli, Yaws, Bejel, Pinta and Hemi-pinta.
Protozoal	Post-kalaazar Dermal Leishmaniasis.
Post-inflammatory	Tinea vesicolor, psoriasis, neurodermatitis, atopic dermatitis, pytyriasis rosea, impetigo, herpes zoster.
Iatrogenic	BAL, Thiouracil and quinacrine.
Miscellaneous	Scieroderma, chronic discoid L. E., sclerocitacrital lichen planus, Lichen albus von Zumbush, Anetoderma and pseudo pelade of Brocq.

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