CLINICOPATHOLOGICAL CORRELATION OF LINEAR DERMATOSES IN CHILDHOOD

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CERTIFICATE

Certified that this dissertation entitled **"CLINICOPATHOLOGICAL CORRELATION OF LINEAR DERMATOSES IN CHILDHOOD"** is a bonafide work done by **Dr.M.CHINNASAMY** Post graduate student of Department of Dermatology, Leprology and Venereology, Stanley Medical College, Chennai - 1, during the academic year 2006 - 2009. This work has not previously formed the basis for the award of any degree or diploma.

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DECLARATION

I **Dr.M.CHINNASAMY**, solemnly declare that the Dissertation titled **"CLINICOPATHOLOGICAL CORRELATION OF LINEAR DERMATOSES IN CHILDHOOD"** is a bonafide work done by me during 2006 - 2009 under the guidance and supervision of **Prof. Dr.M.S.SRINIVASAN, M.D., D.D.,** Professor and Head of the Department of Dermatology, Stanley Medical College, Chennai - 1.

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CONTENTS

INTRODUCTION

Skin is a miracle garment. It is soft, pliable, strong, water proof and self-repairing.

It is the largest organ of the integumentary system, made up of multiple layers of epithelial tissues, and guards the underlying muscles, bones, ligaments and internal organs.

It covers between 1.5 and 2m^2, comprising about one sixth of total body weight, also it is the only organ which is visible and is in direct contact with the environment.

In the examination of the skin, the morphology of individual lesions, their overall pattern and spatial relationship to each other, and their body site distribution are helpful and provide an easily recognizable clue to a rapid visual diagnosis. Indeed, clinical diagnosis is more precise than laboratory tests in many disorders.

Skin lesions present with innumerable patterns like Discoid, Petaloid, Arcuate, Annular, Polycyclic, Livedo, Reticulate, Target, Stellate, Digitate, Linear, Serpiginous, Whorled, etc.

Among these patterns, Linearity is a stellar pattern which attracts the attention of patients and clinicians alike. A single lesion may assume a linear shape or a number of lesions may be arranged in a linear pattern.

The mechanisms or anatomical factors dictating the Linearity are of the following groups.

1. Linear configurations determined by the course of blood vessels, lymphatics or nerve trunks.

2. Linear lesions of developmental origin.

3. Linear lesions following Dermatomal pattern.

4. Linear lesions caused by External factors like Plants, Allergens, Chemicals, thermal and Physical factors (includes Koebner's phenomenon).

5. Linear configurations due to other determinants.

Most of the Linear lesions follow the Blaschko's lines. Children with linear lesions attending the Dermatology Out Patient Department at Govt. Stanley Hospital comprise this study group.

REVIEW OF LITERATURE

HISTORICAL ASPECTS[1-17]

ALFRED BLASCHKO (1858-1922) private practitioner of dermatology in Berlin, whose interest ranged from leprosy to occupational skin disease. He presented his findings on distribution patterns of linear skin disorder at the German Dermatological Society meeting in Breslau in the year 1901[1,2]. He examined more than 140 patients with linear lesions such as epidermal naevi, sebaceous naevi and nevus lipomatosus and carefully transposed the pattern in each patient on to dolls and statues[1,2]. A composite diagram of these distribution patterns was then drawn that has subsequently been referred to as the lines of Blaschko.

BLASCHKO'S LINES defined as, lines which develops following the lines of migration and proliferation of epidermal cells during embryogenesis (i.e., the bands of abnormal skin represent clones of cells carrying a mutation in a gene expressed in the skin).

In 1976, Jackson[2] provided a detailed review of the 1901 publication and introduced the concept of the lines of Blaschko into the English literature, although it has been well known in the European community for decades. These lines do not correspond to other patterns such as Langer's lines of cleavage[3], Voigt's lines (borders between areas

of innervations by peripheral cutaneous nerves[4]), Embroyonic clefts[5], Pigmentary demarcation lines[6], Lines of lymphatic drainage or blood supply.

Although the distribution is linear, the curvature of the lesions does not support the hypothesis that these lines represent Koebner's phenomenon. Most commonly, Blaschko's lines are confused with dermatomes, the segments of skin that are defined by sensory innervation[7]. A major reason for these confusion is that both distribution patterns are characterized by a striking demarcation of cutaneous lesions at the midline. As a reflection of these confusion, several diseases that follow Blaschko's lines are referred to as dermatomal or zosteriform, for example, zosterifom Porokeratoses, zosteriform Lichen planus and zosteriform Lentiginous naevi.

The Blaschko's lines were most apparent on the trunk with arcs on the upper chest, S-shape on the abdomen, a V-shape as the lesions approach the posterior midline and spirals on the scalp are seen.

Occasionally, however, the lesions of herpes zoster do appear to have a more figurate arrangement, raising the possibility that the migration of cutaneous nerves may influence the pattern of Blaschko's lines. On the lateral foot, the lines of Blaschko respect the junction between plantar skin and hair bearing skin and therefore overlap with Wallace's line[8].

The linear arrangement of the lines of Blaschko's points to a relation with metamerism (body structures that exhibit a series of segments), and the possibility has been raised that these lines represent the distribution of the autonomic innervations to the dermal viscera, that is the visceral afferents, as opposed to the sensory afferents (Edmund S, Crelin, Ph.D., Personal Communication, June 1992). The possibility has also been raised that Blaschko's lines simply represent stretching of the skin during embryogenesis, an analogy given in the pattern seen when newspaper print is superimposed on Silly Puty, then stretched (Lawrence Solomon, MD, Personal Communication, July or August 1993). However, involvement of fat and blood vessels is difficult to explain with this theory[9].

Although the lines of Blaschko clearly do not correspond to the distribution of hair tracts, the possibility of overlap with hair whorls has been raised. This debate is, in part, based on the fact that those lines of Blaschko are less well-defined on the head and neck. Happle et al[10,11] have added lines to the posterior scalp, whereas they have attempted to delineate further the lines on the lateral aspect of the face and neck. Brown and Gorlin[12] reviewed the literature in 1960 and mentioned vertical striations in the lips, linear midline lesions on the hard and soft palate, and linear unilateral and / or midline bands on the tongue in patients with epidermal naevi.

The anatomic equivalent of Blaschko's lines has been described in the eyes as well as in teeth. For example, female carriers of X-linked ocular Albinism can have a striated pigmentary pattern in the peripheral retina[13] in addition to an alternating spokewheel - like pigmentation of the iris (alternating normal pigmentation and hypopigmentation[14]). Female carriers of X-linked cataracts and X-linked Lowe's (oculocerebrorenal) syndrome have sectorial cataracts and lens opacities with an irregularly radiated pattern[15]. Of note, similar sectorial cataracts and radial patterns in the lens have been described in women with X-linked dominant chondrodysplasia punctata[16]. Witkop[17], also described alternating vertical bands of opaque white and translucent (normal-appearing) enamel on the central incisors of women heterozygous for X-linked hypomaturation Amelogenesis imperfecta.

MOSAICISM[17,18,19]

The distribution of cutaneous lesions implies the presence of two different clones of cells in early embryogenesis. The various explanations for the clones include Lyonization in X-linked disorders, Post zygotic somatic mutations in sporadic conditions and gametic half-chromatid mutations[17].

Mosaicism describes an individual with two or more cell lines of different genotypes derived from the same zygote. In health, all females exhibit functional mosaicism with regard to their X chromosomes. One

of the two chromosomes in the cells of normal females undergoes inactivation at an early stage of embryonic development (12-16 days after fertilization), a process known as LYONIZATION.

In 1961, Mary Lyon reported striped patterns for some X-linked color genes in mice. She hypothesized that the stripes reflect two populations of cells, one expressing maternal X chromosome and the other paternal. In 1965, Curth and Warburton[18] applied the Lyon hypothesis to the X-linked Incontinentia Pigmenti which is characterized by lesions following Blaschko's lines.

In 1977, Happle[19] recognized lyonization as the cause of Blaschko's lines in female patients heterozygous for other X-linked disorders. Blaschko's lines have been comprehensively reviewed by Bolognia et al, and cutaneous mosaicism more recently by Paller.

CHIMERISM[18]

It denotes the presence of two or more genetically distinct cell population in an individual derived from two different zygotes.

CAUSES OF MOSAICISM AND CORRESPONDING PATHOGENSIS[20]

CAUSES OF MOSAICISM	PATHOGENESIS
Lyonization	The hypothesis, proposed by Mary Lyon, states that only one X chromosome is active in each female cell, with the other forming the Barr body. Whether the paternal or maternal X chromosome is inactivated is random, but once the choice has been made it is the same in all daughter cells.
Post zygotic (somatic) Mutation	A mutation occurring after fertilization.
Chimerism	Fertilization of one egg by two sperms, or fusion two zygotes, resulting in an individual composed of two genetically different cell lines.
Chromosomal non-disjunction	The failure chromosome to separate correctly during either meiosis or mitosis, resulting in daughter cells with aberrations of chromosome number or structure.
Half Chromatid Mutation	A mistake in DNA polymerization during the first meiotic division of gametogenesis, whereby the wrong base is synthesized at one point, resulting in a mismatched double strand. If this mismatched chromosome is passed on to the next generation, the first time it separates in mitosis it will provide two templates that are not exactly complementary, giving rise to two different lines of daughter cells.

MOSAICISM - THE DIFFERENT PATTERNS[21]

TYPE 1 : LINES OF BLASCHKO

Fountain like pattern	-	back
S-figure	-	lateral and ventral aspect of trunk
Spiral	-	scalp

These lines reflect the dorsoventral outgrowth of embryonic cells from the neural crest. Their proliferation interfere with the longitudinal growth and increasing flexion of the embryo, resulting in a characteristic arrangement.

Head and neck - variable pattern, tent to intersect at an angle of 90^{o}

TYPE 1.a - narrow bands (e.g.: Incontinentia pigmenti)

TYPE 1.b - broad bands (e.g.:MeCune-Albright syndrome).

TYPE 2 : CHECKBOARD PATTERN

Flag like area with a sharp midline separation (distributed in a random way and not alternating regularly).

E.g. : Speckled lentiginous naevus, Becker nevus

Pattern of patchy hairiness as noted in women heterozygous for X-linked hypertrichosis.

TYPE 3: PHYLLOID PATTERN

Leaf - like patches and oblong macules (midline separation is not always present).

E.g.: Phylloid hypomelanosis (neurocutaneous syndrome)

Phylloid pattern of hyperpigmentation

TYPE 4: LARGE PATCHES WITHOUT MIDLINE SEPARATION

E.g.: Congenital giant melanocytic nevi, with or without neurological Involvement (clonal origin)

Acquired melanocytic nevi.

TYPE 5: LATERALIZATION PATTERN

E.g.: CHILD syndrome (X-linked dominant, male lethal-trait)

CHILD nevus - one half of the body, with a sharp midline demarcation.

X-inactivation coincides with the origin of a clone of organizer cells controlling a large developmental field.

ZOSTERIFORM NEVI[21]

A zosteriform arrangement corresponds to the system of dermatomes but all nevi are dermatomal but follow the lines of Blaschko.

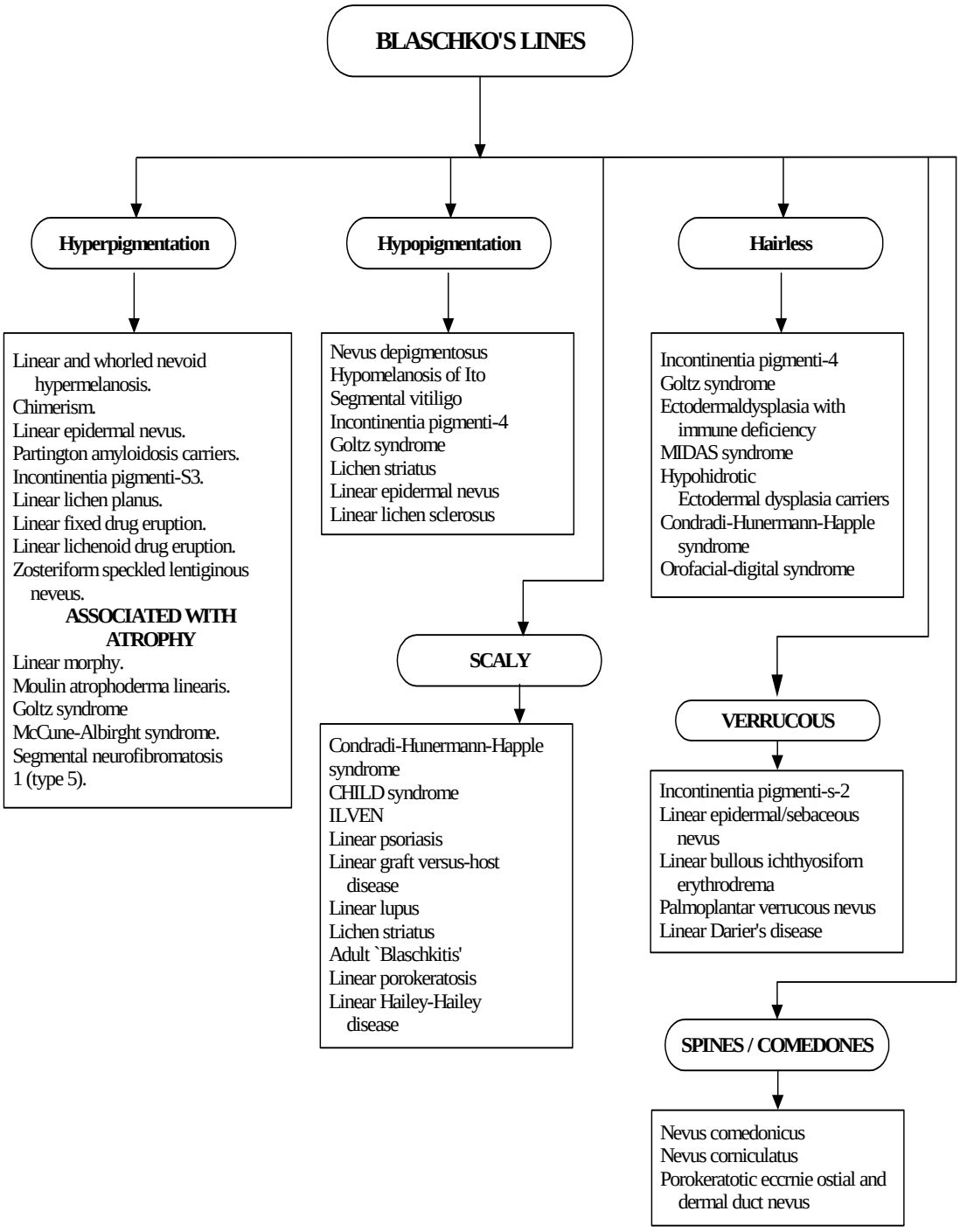
LINES THAT DO NOT INVOLVE MOSAICISM[21]

1. Lines of Voigt-boundaries of peripheral cutaneous innervations.
2. Matsumoto line (also Futcher's line) pigment demarcation line on arms and legs.
3. Meridian lines of Acupuncture.

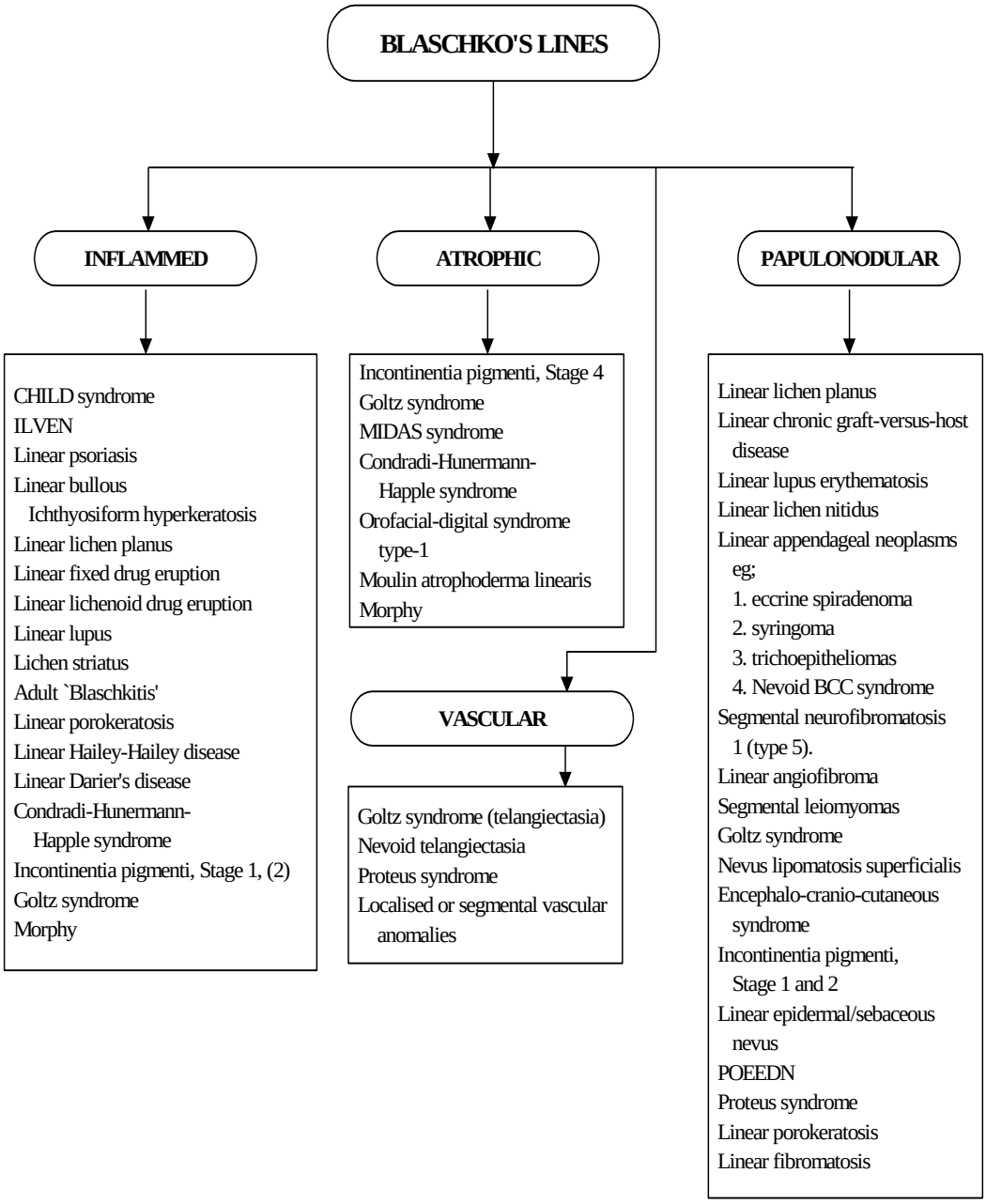
ANATOMICAL AND CAUSATIVE FACTORS IN LINEAR LESIONS[23]

Blood vessels	-	Thrombophlebitis, Mondor's disease
	-	Eczema related to varicose veins
	-	Temporal arteritis
Lymphatics	-	Lymphangitis
	-	Sporotrichosis, Fish tank granuloma
Dermatome	-	Herpes zoster, zosteriform naevus, zosteriform Darier's disease,
	-	Zosteriform metastases
Nerve trunks	-	Leprosy (thickened cutaneous nerves)
Developmental	-	Pigmentary demarcation line, linea nigra
(Blaschko lines)	-	Epidermal naevi, Incontinentia pigmenti, Lichen striatus, Hypomelanosis of Ito, Linear Psoriasis, Linear Lichen planus.
Skin stretching	-	Striae due to growth spurt (on lower back)
Infestation	-	Scabies, Larva migrans (serpiginous)
External factors	-	
Plants	-	Phytophotodermatitis
Allergens	-	Elastoplast, nail varnish (neck), necklace
Chemicals	-	Caustics, eg. Phenol
Thermal	-	Burns
Physical	-	*Trauma to previous normal skin* Keloid scar, bruising, dermatitis artifacta, Amniotic constriction bands *Trauma to skin with a pre-existing dermatosis* Purpura (cryoglobulinaemia, amyloid, vasculitis) Blisters (epidermolysis bullosa, porphyria) *Koebner phenomenon* Psoriasis, Lichen planus, Lichen nitidus, Vitiligo, Lichen sclerosus, Pityriasis rubra pilaris. *Inoculation* : Molluscum contagiosum *Other mechanism* : Scar sarcoid
Other determinants	-	Linear scleroderma (limb, forehead)
	-	Senear-Caro ridge (on hands in psoriasis)
	-	Dermatomyositis (dorsum of fingers)
	-	Flagellate pigmentation due to cytotoxic drug

LINEAR CUTANEOUS LESIONS THAT CAN FOLLOW BLASCHKO'S LINES[22]



LINEAR CUTANEOUS LESIONS THAT CAN FOLLOW[22] BLASCHKO'S LINES (Conti...)



LICHEN STRIATUS[25,26]

(Variants of this disorder has also been called blaschkitis, Blaschko linear acquired inflammatory skin eruption, zonal dermatosis, linear neurodermatitis, linear dermatosis, linear lichenoid dermatosis, lichenoid eruption, systematized lichenification and linear eczema).

Lichen striatus is an inflammatory papular eruption with a distinctive linear distribution, often following Blaschko's line, which should be differentiated from many other cutaneous disease with linear pattern[25,26].

DEFINITION[25,26,27]

Lichen striatus is an uncommon self limiting linear dermatosis with unknown aetiology and spontaneous regression. It primarily occurs in children from 5-15 years of age. The average age at diagnosis is 3 years[25,26]. It may also be seen in adults. Cases in two extremes of age have been reported. Females are affected more than males. Females are affected 2 or 3 times as frequently as males[27].

AETIOLOGY[28-32,40]

The exact etiology of the condition is unknown although case clustering and spring / summer preponderance raises the possibility of an environmental or infective basis[28,29].

Seasonal occurrence and simultaneous involvement of siblings has suggested an infectious cause, perhaps viral[28]. Simultaneous occurrence of lichen striatus in two pairs of siblings : Two sisters who had lichen striatus at an interval of six months and, a brother and sister who had the dermatosis contemporaneously after an episode of flue like fever. In all four patients family history was positive for atopy. Lichen striatus frequently associated with atopic diseases, the abnormal immune status of patients with atopy may be predisposing factor in the induction of lichen striatus.

The simultaneous occurrence of lichen striatus in siblings, after a flue like fever appears to corroborate the hypothesis that a viral infection is a possible candidate, as other authors have proposed. The variety of familial cases of lichen striatus is, due to the exceptional confluence of difference sporadic events (atopy, viral infection caught at a specific period of life such as childhood, and a genetic predisposition), simultaneously present in the same patient[40].

The development of lesion along Balschko's lines raises the possibility of a cell-mediated autoimmune reaction to an abnormal clone of cells. Blaschko's lines are believed to represent the direction along which epidermal growth centers expand during early skin development[30]. It has been suggested that the distribution of lesions in lichen striatus may reflect a post zygotic abnormality such as somatic

mutation affection localized stem cells[30]. Manifestation of atopy with abnormal immune response[31]. About 80% of patients have the family history of atopy[32].

PATHOGENESIS[33,34]

In lichen striatus it has been found that the inflammatory cells reaching the epidermis are CD8+ (Suppressor-cytotoxic) T-lymphocytes[33] with the Langerhans cells population in the epidermis either decreased or increased. These findings suggest a cell-mediated immunologic mechanism where cytotoxic events against kerzatinocytes could be taking place during the evolution of the disease[33].

Immunohistochemistry has shown that most of lymphocytes in the upper dermis and epidermis are CD7+, and most of the lymphocytes in the epidermis are CD8+ T-cells expressing HLA-DR+ antigen on their surface[34]. These findings suggest a cell-mediated immunologic mechanism. In one study CDla Langerhans' cells were either decreased or increased or normal in the epidermis[34].

CLINICAL FEATURES[25-29, 35-39]

The morphology of the lesions is distinct. Lesions appear as small, pink, lichenoid papules which are at first discrete, but coalesce rapidly into plaques following Blaschko's lines[25,26]. The lesions start

suddenly and extend over the course of a week or more to become dull red slightly scaly bands.

The width is usually 2mm to 2cm and is often irregular. The bands may broaden into plaques. The length may vary from few centimeters to several centimeters, or may extend the entire length of the limb. The bands may be continuous, interrupted, parallel or zosteriform[25,26].

The lesions occur commonly on one arm or leg or on the neck, but may develop on the trunk, abdomen, buttocks or thighs[25,26]. Rarely the lesions may be multiple and bilateral[27,28]. The lesions are normally asymptomatic, but pruritus of moderate to severe degree may be experienced[29].

Variations - verrucous lesions with confluence (by Johnson)[35]

Light to yellow coloured grouped papules (by netherton)[36]

Flat topped papules (by Frainbell)[37]

Papules, vesicles and crusting (by Felix pinkus)[38]

Hypo pigmentation may be prominent, especially in dark skinned persons. In pigmented skin, post inflammatory hypo pigmentation is a useful sign for distinguishing lichen striatus from linear lichen planus. When lesions extend to the ends of digits, nail involvement may range from fraying to total nail loss[39].

DIFFERENTIAL DIAGNOSES[39]

The differential diagnosis of lichen striatus include linear lichen planus, linear lichen nitidus, linear epidermal nevus, inflammatory linear verrucous epidermal nevus, linear psoriasis and linear lichen simplex chronicus, linear Porokeratosis and linear Dariers disease.

HISTOPATHOLOGY[30, 32-34, 41, 42]

Although lichen striatus has been recognized by its variable histologic picture, some constant microscopic findings may be present. There is usually a superficial perivascular inflammatory infiltrate of lymphocytes admixed with a variable number of histiocytes. Plasma cells and eosinophils are rarely seen[34]. Focally, in the papillary dermis the infiltrate may have a band like distribution with extension into the lower portion of the epidermis, where there is vascular alteration of the basal layer and necrotic keratinocytes. In these areas, the papillary dermis occasionally contains melanophages[30,32]. Additional epidermal changes consist of spongiosis and intracellular edema often associated with exocytosis of lymphocytes and focal parakeratosis.

Less frequently, there are scattered necrotic keratinocytes in the spinous layer as well as subcorneal spongiotic vesicles filled with Langerhans cells[30,33]. A very distinctive feature is the presence of inflammatory infiltrate in the reticular dermis around hair follicles and

eccrine glands. Zang and Mcnutt found combining features of sweatgland (or) hair follicle involvement with other histological features[42]. An unusual perforating variant of lichen striatus has been described, which shows Trans epidermal elimination of clusters of necrotic keratinocytes.

TREATMENT[42]

Because the lesions are self-limited and resolve spontaneously within one year usually there is no treatment necessary. Reassuring the patient is essential. Post inflammatory hypo pigmentation may persist longer. Topical cortico steroids may cause more rapid involution of the erythematous lesions and control of pruritus.

PEDIATRIC BLASCHKITIS[43-45]

(BLASCHKO LINEAR ACQUIRED INFLAMMATORY SKIN ERUPTION; BLAISE)

Expanding the spectrum of childhood acquired Blaschko-linear dermatoses.

Of late, there have been reports of relapsing, pruritic, papulovesicular eruptions in multiple bands, along Blaschko lines on the neck, trunk, and extremities of pediatric age group patients.

Skin specimens in those patients revealed spongiotic dermatitis. This represents the first report of “blaschkitis” in children, providing

further evidence that lichen striatus and blaschkitis are related acquired Blaschko-linear dermatoses that exist on a spectrum rather than as the childhood and adult form of a single disease entity[43]. It would be difficult to distinguish from linear Grover's disease[44]. Taieb et al[45]., considered that 'adult Blaschkitis' represents and adult version of lichen striatus, and proposed the acronym BLAISE to cover both. BLAISE should perhaps be regarded as a description rather than a diagnosis, useful category for many of the disorders in this section, pending more precise identification.

CHILDHOOD LINEAR PSORIASIS[46-49]

Linear distribution of psoriasis is a very rare form. The psoriatic lesion presents as linear lesion most commonly on the limbs, but may also be limited to a dermatome on the trunk.

This may be an underlying nevus, possibly an ILVEN, as these lesions resemble linear psoriasis both clinically and histologically.

This presentation offers to physicians some diagnostic difficulty, especially in the absence of a history of pre-existing psoriasis (or) in the presence of any other linear dermatosis.

Three cases of girls, ages 4 years, 5 years and 10 years with linear psoriasis have been reported.

The clinical features and differential diagnosis of this skin disease which, in children can be easily mistaken for ILVEN.

Atleast three clinical entities have been described as “linear psoriasis”[49]. All follow Blaschko’s lines. The first, and most common, is an ILVEN that may resemble psoriasis clinically. Since Woringer’s first report[46] in 1936, the psoriasiform nature of some epidermal nevi has been recognized[47]. These lesions often exhibit a characteristic histologic picture of areas of hypergranulosis and orthokeratosis alternating with agranulosis and parakeratosis, although features of psoriasis may also be present.

A second entity appears to present the extension of psoriasis into an epidermal nevus by the isomorphic (Koebner’s) phenomenon. Patients may exhibit or develop typical lesions of psoriasis outside the segmental area involved by the nevus[48,49].

The existence of third true “linear psoriasis” that does not fit into other types is controversial. Nonetheless, at least two children have been described with extensive lesions following Blaschko’s lines that resembled psoriasis both clinically and histologically[49].

In neither case was a pre-existing nevus present, nor were there any signs of psoriasis elsewhere on the patient’s skin. It has been

suggested that psoriasis in such patients arises by somatic recombination, giving rise to the pattern following Blaschko's lines.

The lesions could be distinguished from invasion of a verrucous epidermal nevus by psoriasis as a result of the isomorphic phenomenon and from dermatitic epidermal naevi, by their minimal pruritus and their therapeutic response to ultraviolet radiation. Linear psoriasis is easily confused with ILVEN[47].

LINEAR LICHEN PLANUS[50-56]

Lichen planus is a papulosquamous disorder with an insidious onset in most cases. It can occur in families and there is a genetic predisposition, as reported in monozygotic twins. An increased frequency of lichen planus is noted with HLA-B7, HLA-28, HLA-DR1 and HLA-DR10.

The morphologic variants of cutaneous lichen planus seen in pediatric patients are linear, hypertrophic, annular, follicular, erosive, actinic and bullous. Linear lichen planus is the most common variant seen in pediatric age group. It is characterised by typical papules arranged in a linear or zosteriform pattern on the limbs and less commonly, the trunk. It is distinct from linear lichen planus that develops at sites of trauma (Koebner phenomenon)[50-52].

Linear lichen planus was first described by Devergie in 1854 and accounts for 0.24-0.62% of all the patients with lichen planus and was found to be more common in Japan[53].

Scattered linear lesions often occur in patients with lichen planus and are a result of a combination of scratching and the Koebner's phenomenon. Less commonly, unilateral streaks or bands of LP are seen that are longer and wider than the trauma induced lesions[49]. In a review of large series of pediatric lichen planus cases a predliction for male gender was revealed, the age of onset being from the first year through to adolescence[50-52,54]. More than half the children had pruritus.

In the majority of cases, the streaks were composed of polygonal violaceous papules and coalescing plaques. Surface of lesions shows Wichkam's striae (white lines). It mainly occurs on the front of wrists, lumbar region, around the ankles and glans penis. If lesions extend to the end of digit there may be associated nail dystrophy. Palms, soles, nails and oral mucous membranes may also affected.

Multiple linear lesions following the lines of Blaschko have been reported in lichen planus. They may occur as isolated, long, narrow, linear bands extending the whole length of the limb at times and more common in childhood[53].

Histopathology of a typical papule of lichen planus shows compact orthokeratosis, wedge shaped hypergranulosis contributing clinically to wichkam's striae, irregular acanthosis and pointed lower ends of the rete ridges, giving them a 'saw - toothed' appearance. Liquefactive degeneration of the basal layer, with formation of civatte or colloid bodies. Dense band like inflammatory infiltrate composed of lymphocytes and histiocytes, closely hugging the lower end of the epidermis and also present perivascularly are present. Melanophages are seen in the upper dermis. Lichen planus is an immunologically mediated dermatosis as evidenced by the immunofluorescence studies[55].

- Treatment includes, moderately potent or super potent topical steroids are the treatment of choice for cutaneous lichen planus in pediatric patients.

- Close supervision of potent topical steroids therapy is required to minimize the risk of developing local side effects.

- Topical steroids can be combined with oral corticosteroids (0.5 to 1.0mg / kg per day), administered as a tapering dose over a 2-12 weeks period.

- In acute eruptions and lesions of scalp, nail and oral mucosa, systemic steroids (prednisolone) 15-20mg are given daily for 6 weeks and then gradually tapered off.

- Oral acitretin in severe cutaneous lesions are effective.

- Low dose Tretinoin was found to be effective and well tolerated in lichen planus refractory to other treatments[56].

- Despite lack of evidence of proven efficacy, cyclosporin, Griseofulvin, puvaphototherapy dapsone and phenytoin have been used with difficult patients not responding to steroid and oral retinoids.

- Most pediatric patients with lichen planus responds to treatment with full clearance over 1-6 months.

EPIDERMAL NAEVI[21,57]

(VERRUCOUS NEVUS, NEVUS UNIUS LATERIS)

Epidermal nevus is a developmental malformation of the epidermis in which an excess of keratinocytes, sometimes showing abnormal maturation, results in a visible lesion with a variety of clinical and histopathological patterns[57]. Epidermal nevus may present either as keratinocytic epidermal nevus, or it may have differentiation towards sweat gland, sebaceous gland or hair follicle.

Verrucous epidermal nevus is a congenital, non inflammatory cutaneous hamartomas composed of keratinocytes. They are divided into epidermolytic and non-epidermolytic type[21].

Verrucous epidermal nevus consist of hyperplasia of the surface epidermis and typically appear as verrucous plaques that coalesce to form well-demarcated, skin coloured to brown, papillomatous plaques. Most lesions are present at birth or develop during early infancy; they enlarge slowly during childhood and generally reach a stable size at adolescence. Lesions may be localized or diffuse. Linear configurations are common, especially on the limbs, and may follow skin tension lines, or Blaschko's lines. If plaques are minimally elevated and multiple, can be mistaken for Linear and Whorled Nevoid Hypermelanosis.

Epidermal nevi can involve the palms and soles as well as the oral mucosa. When it occurs over the palm, it may be confused with linear keratoderma. Additional findings include Wooly hair nevus, straight hair nevus and nail dystrophy.

HISTOPATHOLOGY[58]

Hyperkeratosis, papillomatosis and acanthosis with elongation of rete ridges. Epidermolytic hyperkeratosis or focal acantholytic dyskeratosis[58]. The salient histological features of epidermolytic hyperkeratosis are A) Perinuclear vacuolization of the cells in the

stratum spinosum and in the stratum granulosum. B) Peripheral to the vacuolization irregular cellular boundaries. C) An increased number of irregularly shaped, large keratohyaline granules and. D) Compact hyperkeratosis in the stratum corneum.

INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS (ILVEN; DERMATITIS EPIDERMAL NEVUS)[58,59]

Altman and Mehregan[59] coined the phrase “ILVEN” to describe a sbuset of epidermal nevi that were erythematous, inflamed and pruritic. These nevi comprise a unique variety of keratinocytic epidermal nevus which exhibit both psoriasiform and inflammatory features. These nevi follow Blaschko’s lines.

Etiology include : mosaicism of a dominant mutation, partial expression of Retroposons, clonal dysregulation of growth triggered by HIV infection, absence of involucrin expression in epidermis[58].

Diagnostic criteria include the following[59]:

Early age of onset

Female predominance (4:1)

Frequent left lower extremity involvement

Pruritus

Classical biopsy finding and

Lesional persistence with refractoriness

PATHOGENSIS[59,60]

The lesions occur primarily on the lower extremities, with a slight preference for the left leg. Sometimes multiple Blaschko-distributed linear verrucous plaques most commonly on legs, pelvis and buttock may be seen. Although the Involucrin expression has a very characteristic pattern in ILVEN. The orthokeratotic epidermis shows almost negative staining for involucrin is expressed prematurely in most of the suprabasal keratinocytes[59].

More than 90% of the mononuclear cells in the dermal infiltrate are CD4+ (helper-inducer) T-lyphocytes; in contrast, may be present at birth, the majority of ILVEN appear during infancy and childhood. Fifty percent are evident by 6 months of life and 75% by 5 years of age. Histologic examination of ILVEN demonstrates psoriasiform hyperplasia of the epidermis and alternating parakeratosis without a granular layer and orthokeratosis with a thickened granular layer. Underneath the parakeratotic areas, there is mild exocytosis of lymphocytes and slight spongiosis. The papillary dermis shows a mild to moderate perivascular inflammatory infiltrate of lymphocytes and histiocytes[60].

Clinically ILVEN are stable and do not respond to UV light or topical medications. Treatment of ILVEN is often challenging. The presence of overlapping psoriasis component will respond to

antipsoriatic treatment. Other cases have improved with topical steroids and topical retionoids, oral retinoids, and destructive modalities such as excision, ablative laser, cryotherapy, and dermabrasion. Surgical excision is an option, but the risk of scarring needs to be considered. Unlike the non-inflammatory epidermal nevi, ILVEN is not associated with neurological defects.

Rarely, there are Ipsilateral skeletal abnormalities, usually reduction deformities. But these abnormalities may instead represent examples of CHILD or Conradi-Hunermann syndromes. An alternative acronym that has been used to describe this association is PENCIL (Psoriosiform Epidermal Nevus with Congenital Ipsilateral Limb defects).

TYPE OF EPIDERMAL NEVUS (described by Solomon and Esterly)[61]

1. Nevus Unius Lateralis - most common type
2. Ichthyosis hystrix
3. Acantholytic Epidermal Nevus
4. Linear Nevus Sebaceous
5. Localized Linear verrucous nevus
6. Velvety Epidermal Nevus of Axilla
7. Mixed type

EPIDERMAL NAEVUS SYNDROME[61,67]

(Solomon syndrome, Sebaceous Naevus syndrome; Schimmel Penning's syndrome; Feverstein-mims syndrome; Organoid Naevus syndrome; Jadassohn's Naevus phakomatosis)

Epidermal nevus syndrome is a rare condition that refers to the association of epidermal nevi with abnormalities in other organ system including nervous, skeletal, eye, cardiovascular and urogenital system. Although most cases occur sporadically, AD transmission has been reported as well. Both sexes are equally affected and age of diagnosis ranges from birth to 40 years[67].

ETIOLOGY AND PATHOGENSIS[67]

Epidermal nevi are organoid nevi that arise from pluripotent germinative cells in the basal layer of the embryonic epidermis. Histological variation is common with different cell components seen in different areas of the same lesion and hence Solomon and Esterly the term *epidermal nevus* to encompass all the variants.

Epidermal nevus appears to be a form a genetic mosaicism. The concept of AD lethal gene surviving in only mosaic state was proposed by Happle to explain the genetic basis of several syndromes under this condition. Approximately 5-10% of Epidermal nevus may show features

of Epidermolytic hyperkeratosis which is a Forme fruste of Bullous ichthyosiform erythroderma.

CLINICAL FEATURES

Most Epidermal nevi are isolated and can occur at any site but usually do not involve the head and neck but commonly present in the these areas as Nevus Sebaceous of Jadassohn. Lesions are generally found along Blaschko lines and seldom cross the midline. Whorled patterns may also be seen.

Apart from this Epidermal nevi, when extensive should arouse the suspicion of Epidermal nevus syndrome. In over 80% of cases the onset is within one year of age. The lesions extend beyond their original distribution but reach stability in late adolescence without any further progression.

SYSTEMIC ABNORMALITIES[62]

1. Skeletal - Bone deformities, cysts, atrophies and hypertrophies.

2. Neurological - Mental retardation, seizures (due to hydrocephalus, cerebrovascular malformation and intracranial calcification) and ocular defect.

3. Risk of Malignancy - generally limited to Nevus Sebaceous of Jadassohn. Visceral malignant associations include Wilms tumor, Nephroblastoma, adenocarcinoma of salivary glands / esophagus and stomach.

DIFFERENTIAL DIAGNOSIS

1. Schimmel penning syndrome - sebaceous nevi associated with cerebral abnormalities, coloboma and conjunctival lipodermoid.

2. Nevus comedonicus syndrome - cataracts, skeletal defects and ECG abnormalities.

3. Pigmented hair epidermal nevus syndrome - Becker’s nevus, Ipsilateral breast hypoplasia and skeletal defects.

4. Proteus syndrome - soft flat epidermal nevi, limb gigantisms, skeletal hyperplasia and subcutaenous Hamartomas.

5. CHILD syndrome.

6. Phakomatosis pigmentokeratotica - sebaceous nevus, contralateral speckled lentiginous nevus.

The importance of this syndrome is screening of patients for associated systemic manifestations. The epidermolytic pattern of histology is a mosaic pattern due to keratin 1 and 10 involvement. Genetic counseling is required for the Epidermolytic type.

NEVUS SEBACEOUS (OF JADASSOHN)[63-66]

Sebaceous nevi are present in approximately 0.3% of newborns and appear as a waxy to verrucous plaques[63]. Typically, there is a yellow to orange hue that reflects hyperplasia of sebaceous glands. Most commonly over head and neck but also on extremities and trunk[64]. Distribution in along lines of Blaschko, but difficult to appreciate on scalp, face or neck. It is always present at birth, and is typically a pink-yellow (or) yellow-orange plaque. Frequently recognised as a patch of localized atopecia[65].

In sebaceous nevus syndrome (Schimmelpenning-Feuerstein-Mims syndrome), patients have ocular, vascular, musculoskeletal and CNS abnormatlities in addition to sebaceous nevi.

Ocular : epidermal nevus involving the eyelid (or) conjuntiva, choristomas, cortical blindness, microphthalmia, macrophthalmia, anophthalmia, corneal opacities and cataracts. Colobomas and lipodermoid tumors of conjunctiva or sclera[66].

CNS : seizures, mental retardation and intracranial Hamartomas.

Skeletal : hypophosphatemic osteomalacia / rickets.

HISTOPATHOLOGY[68]

Increased number of sebaceous glands, apocrine sweat glands, in the dermis, absent or hypoplastic hair follicles and papillomatosis of the epidermis. Cords of undifferentiated cells resembling the embryonic stage of hair follicle are present in the dermis.

LINEAR MORPHOEA AND PARRY-ROMBERG SYNDROME[70-75]

- Morphoea is a connective tissue disorder of unknown etiology, characterized by sclerosis of the skin and subcutaneous tissues and it is an uncommon disease.

- Schachner's analysis of referrals estimate that only 3 of 1578 patients (0.19%) seen over a two year period had morphoea[69].

- In another report of 68 children were diagnosed with morphoea over a 30 year period, this represents 0.18% of dermatologic patients seen[70].

- Of the 68 cases, 26 were linear 38.2%, 15 were en coup de sabre (22.1%), 14 cases were plaque type (20.6%), nine had generalised morphoea (13.2%) and four had the pansclerotic form (5.9%).

The morphoea can follow the lines of Blaschko. Trauma and immobilization may be provocative factors. It has also occurred following BCG vaccination[71], Varicella, injections of vitamin K and at the site of radiotherapy. Hormonal factors may influence as observed by exacerbation or development during pregnancy. It may occur after measles or infection with Borrelia burgdorferi[72]. Familial incidence is noticed. Frontal type of Morphoea appears to have a genetic basis.

It may be associated with phenylketonuria and has occurred in those on treatment with penicillamine and bromocriptin. Autoimmune aetiology is incriminated as evidenced by increased incidence of organ-specific auto antibodies in patients and relatives and associated with idiopathic thrombocytopenic purpura. Females are 3 times more affected than males. Peak incidence is between 20 and 30 years of age group, although 15% being below 10 years of age[73].

A childhood onset occurs in 2-3 percent of all cases of Morphoea. The clinical features are essentially similar to adults, however a prior history of trauma is usually present in 25 percent of children. Children are also less likely to show serological abnormalities as compared to adults[74].

Linear morphoea is different from plaque morphoea, with respect to age of onset, distribution, clinical outcome and also serology. Encoup de sabre (sabre hit)[75] is linear morphoea of the forehead. One distinct aspect of linear morphoea is the frequent association with high titers of ANA, including biochemical analysis of an individual lesion is indistinguishable from that of classical plaque morphoea.

Linear morphoea may present initially as a linear erythematous, inflammatory streak, but more frequently it begins as a harmless appearing lesion of plaque-type morphoea that extends longitudinally as a series of plaques that join to form a scar-like band that may severely impair the mobility of the affected limb.

Linear morphoea tends to involve the underlying fascia, muscle and tendons. This leads not only to muscle weakness but shortening of the muscles and fascia impairs joint motility. Linear morphoea is especially dangerous when extending over joints, as this almost invariably results in disabling joint immobilization. If only the subcutaneous tissue, muscle and occasionally bone involved, this ipsilateral form of linear morphoea is known as progressive facial hemiatrophy (or) parry - Romberg syndrome.

HISTOPATHOLOGY[76-87]

i. Early inflammatory stage - found particularly at the violaceous border of enlarging lesions, the reticular dermis shows interstitial lymphoplasmacytic infiltrates among slightly thickened collagen bundles.

ii. Intermediate stage - infiltrates surround eccrine coils. More infiltrate is found in dermis and subcutaneous fat. Large areas of subcutaneous fat are replaced by newly formed collagen, composed of fine, wavy fibers. Endothelial swelling and edema of walls of vessels are seen.

iii. Late sclerotic stage - inflammatory infiltrate disappears and epidermis is normal. Collagen bundles appear thickened, closely packed and hypo cellular. In papillary dermis, collagen appears homogenous. Eccrine glands appear atrophic with few adipocytes surrounding them; blood vessels are fibrotic with a narrowed lumen. Elastic fibers are thickened and arranged parallel to epidermis and collagen fibers.

iv. The fascia shows fibrosis and sclerosis[76].

The en coup de saber types of morphoea represent linear morphoea of the head. It is normally unilateral and extends from the forehead into the frontal scalp. It may start either as a linear streak or a row of small plaques that coalesce. A paramedian location is more common than a median location. Like plaque morphoea, it may initially be surrounded by a discrete lilac ring that extends longitudinally and may reach the eyebrows, nose and even the cheeks. The waning inflammation leaves a linear, hairless crevice that in some patients is more sclerotic while in others is more atrophic.

En coup de sabre morphoea can also involve the underlying muscles and osseous structures. Rarely, the inflammation and sclerosis progress to involve the meninges and even the brain, creating potential focus for seizures. Alternatively, very slowly progressing inflammation, indistinguishable from the inflammatory process of linear morphoea, leads to gradual involution of the skin, fatty tissues and underlying bones.

Laboratory abnormalities may show an elevated ESR, eosinophilia and hypergammaglobulinemia. ANA is positive in 67% of patients with Linear Scleroderma (either homogenous or speckled type)[77]. Anti - U1RNP antibodies may be present[78]. Anti-single stranded antibody and Anti-histone antibody may be present which may indicate

the aggressiveness of the disease[79]. Rheumatoid factor may be positive[80].

Serum concentration of procollagen type - I carboxy terminal propeptide level is a useful indicator of disease severity with localized Scleroderma[81]. Elevated soluble CD23 levels in these sera are a new serological indicator of the severity[82]. Hereditary deficiency of complement factor C2 is reported. 20MHZ B-mode ultrasound scanner, non invasive methods can be used to know the thickening and sclerosis of the skin in monitoring the course and treatment of localized Scleroderma[83].

Various drugs are used in the treatment. D-penicillamine and systemic corticosteroids are beneficial in the active phases of the disease. Phenytoin may help sometimes. Vitamin-D analogues both orally and topically may help, by fibroblast inhibition. Calcipotriol (50 microgm/kg) twice daily topical application at night has shown marked decrease in erythema associated telangiectasia, dyspigmentation and induration by the end of 3 months[84]. Chloroquine or Hydroxychloroquine, Methotrexate have been beneficial in some cases[85]. Cyclophosphamide or Azathioprine is reserved for severe and resistant forms of Morphoea. Plasmapheresis with systemic steroids for morphoea with elevated titres of ANA and anti-ds DNA may be helpful. UVA1 phototherapy and PUVA bath photochemotherapy have also been

tried. Tranilast in case of contractures, an anti-allergic drug, support the concept that mast cells have a role in increasing collagen synthesis in the disease[86].

Active and passive stretching exercises of limbs in Linear Morphoea, surgical release of contracture, plastic surgery for 'En coup de sabre' and tissue expansion for Morphoea of face and scalp are done. Dental treatment is needed if jaw is involved[87].

SEGMENTAL VITILIGO[88,89]

- Vitiligo is another multifactorial disorder that occasionally occur in a segmental distribution[88].
- Unilateral macules in a dermatomal (or) quasi-dermatomal pattern.
- Compared with symmetric vitiligo, the linear type is earlier in onset more commonly occurs in children, less likely to spread to other areas of the body, and less frequently associated with other autoimmune diseases[89] (or) thyroid disease.
- Alteration of neuronal peptides has been implicated in the pathogenesis of this type.

- The lesion tend to be broad bands, patches, or blocks corresponding more to dermatomes than Blaschko's lines, perhaps in keeping with a neuronal pathogenesis. This idea is consistent with mosaicism, as the neuronal abnormality could be mosaic, or alternatively, there could be a Clonal susceptibility of melanocytes to neuronal or other influences.

- Morethan one half of the patients with segmental vitiligo have patches of white hair known as poliosis.

- Segmental vitiligo may be confused with nevus depigmentosus, but the latter is characterized by hypopigmentation (rather than depigmentation), a stable size and shape (relative to the growth of the child), and is present at birth or noted soon thereafter. The presence since birth of a linear band with complete absence of pigment raises the possibility of mosaicism for piebaldism.

LINEAR BENIGN TUMOURS[92]

Syringomas, Trichoepitheliomas, and Eccrine spiradinomas are all heritable ectodermal tumours and all occasionally occur in a linear distribution. Leiomyomas are mesodermal and also occasionally occur in a segmental pattern[92].

LINEAR POROKERATOSIS[90,91]

- Linear porokeratosis is an uncommon variant, presents in early childhood, although congenital presentation have been reported, and usually life long.
- Linear porokeratosis is also a linear lesion composed of multiple annular plaques of typical porokeratosis[90].
- Two distinct clinical variants - The more common presentation consists of a unilateral lesion confined to an extremity following Blaschko's lines.
- In the rare generalised form, multiple lesions affect several extremities and involve the trunk.
- Linear porokeratosis is thought to represent a mosaic form of classic porokeratosis of mibelli, resulting from a postzygotic mutation.
- It has been attributed to allelic loss due to mutation (or) over expression of P53.
- Linear variants have the highest potential for malignant degeneration of all the porokeratoses.
- A rare case of squamous cell carcinoma arising from the linear lesions has been reported[91].

AIMS OF STUDY

1. To study the Incidence of Childhood Linear Dermatoses at the Skin Out Patient Department, Government Stanley Hospital, Stanley Medical College, Chennai, during the period Sep.2006 to August 2008.

2. To study the age and sex distribution.

3. To study the various sites of distribution.

4. To study the symptomatology and predisposing factors.

5. To look for other associated conditions.

6. To study the histopathological pattern.

MATERIALS AND METHODS

This study includes 60 cases of linear dermatoses in childhood. They were assessed clinically and histologically and routine investigations like blood, urine and motion examinations. X-ray tests were done wherever necessary.

Detailed and complete history in all the 60 cases studied was taken. Their address, occupation and socioeconomic status were noted. Special reference regarding the marital status of parents was kept in mind to rule out a genetic basis. Sibling history was taken in all cases to rule out an infectious origin if other siblings were affected by same dermatosis.

Great care was taken to find out associated skin disorders like Alopecia Areata or nail changes. Special importance was given to rule out Koebnerization. Other special findings like Auspitz sign for psoriasis and Wickham's striae for Lichen Planus were noted.

For the cases of Epidermal Nevus Syndrome, opinions of specialty departments like Neurology, Ophthalmology, Oto-Rhino-Laryngology and Dentistry were sought.

All cases of lichen planus were screened for HBsAg.

Complete physical examination was done for each child with special reference to lymphadenopathy, mucosal changes, Hair changes and Nail changes. Palms and Soles were also examined.

Skin biopsy was done for amenable children from the advancing edge of lesions. Biopsy slides were studied with H & E staining.

OBSERVATIONS

INCIDENCE OF LINEAR DERMATOSES

No.of new pediatric cases attending our skin OPD, SMC, Chennai - per day.	30
Total number of new pediatric cases attending our OPD during the period of Sep.2006 to August 2008.	9000
Total cases of Linear childhood Dermatoses during this period.	60
Incidence of Linear Dermatoses in childhood.	0.66%

Among the study group of 60 cases, 52 were asymptomatic and reported for cosmetic reasons.

Moderate to severe itching was the main reason to bring the Lichen Planus Children and few of the Lichen Striatus children to treatment.

Age and Sex distribution in this study of 60 children with Linear lesions are shown in Table - I (n=60)

Age (years)	Male	Female	Total
0-2	4	4	8
3-5	5	5	10
6-8	6	8	14
9-12	14	14	28
Total	**29**	**31**	**60**

Total Male cases = 29

Total Female cases = 31

Female : Male Ratio = 31:29

Most of the linear Dermatoses were in the age group between 6-12 years.

The clinical entities among the study group are shown in

Table - II (n = 60)

Clinical Entities	No. of cases	%
Lichen Striatus	25	41.7
Linear Lichen Planus	15	25
Linear Epidermal Nevus	9	15
Linear morphoea	5	8.3
Linear Vitiligo	4	6.7
Linear Psoriasis	2	3.3
Total	**60**	**100**

Among 60 cases, Lichen Striatus was the most common presentation followed by Linear Lichen Planus and Linear Epidermal Nevus in this study. Family history of similar lesions was not present in any of these patients. Out of the 60 cases Fifty six cases showed a unilateral distribution and only the remaining 4 showed bilateral distribution of lesions in a linear pattern. Fifty four cases had lesions mainly over the extremities corresponding to the lines of Blaschko.

LICHEN STRIATUS

Among the 60 children in this group, twenty five presented with lichen striatus in the age group between 1½ months to 12 years and Male : Female ratio of 2:3.

Age and Sex distribution among patients with lichen striatus

Table - III (n = 25)

Age (years)	Male	Female
0-2	2	4
3-5	2	3
6-8	2	4
9-12	4	4
Total	**10**	**15**

14 out of twenty five children in this group were in the age group between 6 and 12 years. Twenty three children were asymptomatic and mainly came for cosmetic reasons, only two children presented with itching. Eighteen children had hypopigmented macules and papules, and another five children had skin coloured tiny papules and the remaining two had erythematous macules. Twenty out of twenty five cases had an interrupted linear pattern of lesions and in the remaining five cases, lesions were continuous.

Most of these lesions were non scaly and only four cases had mild scaling. The duration of the lesions ranged from 2 week to 2 years, with an average of 3 months. Among these 25 children, six children had 2nd and 3rd degree consanguineous parentage and none of the other family members of these cases were affected with similar problems.

The lesions were present mainly over the extremities and length varying from 6 cms to 20 cms, with a mean length of 15 cms.

Site of involvement in children with lichen striatus

Table - IV (n = 25)

Site	**Right**	**Left**
Upper limb	3	5
Lower limb	3	10
Trunk	3	
Trunk & Upper limb	1	
Total	**25**	

Most of the cases had unilateral distribution predominantly over the left side.

Some of the associated skin conditions seen in these children included Insect bite allergy, Tinea versicolor, Xerosis of hands and feet.

None of them had nail changes or mucous membrane involvement.

Skin biopsy was done for relevant cases. Ten cases showed a chronic dermatitis picture consisting of mild to moderate acanthosis, mild spongiosis, and perivascular lymphohistiocytic infiltrates in the dermis. Three cases showed psoriasiform dermatitis like picture consisting of mild hyperkeratosis, regular acanthosis, and sparse inflammatory infiltrates in the upper dermis and blood vessels with sparse inflammatory cells seen in the mid dermis. Three cases showed lichenoid dermatitis like picture consisting of flaky hyperkeratosis, mild to moderate acanthosis, indistinct dermo-epidermal junction, and subepidermal collections of chronic inflammatory infiltrates.

LINEAR LICHEN PLANUS

In this study group fifteen children presented with Linear lichen planus. The age group ranged between 12 months and 12 years with an average of 10 years.

Out of the 15 cases, eight cases were males and seven were females forming a Male : Female ratio of 8:7.

Age and Sex distribution in children with linear lichen planus

Table - V (n = 15)

Age group	Male	Female
0-2	1	0
3-5	1	1
6-8	0	2
9-12	6	4
Total	**8**	**7**

Four out of the fifteen cases, presented with itching with duration of symptom ranging from 1 week to 3 months.

Four children were treated for chronic suppurative otitis media prior to the onset of lesions. All these patients had hyperpigmented, discrete, flat topped papules and plaques of size varying from 0.5 - 1cm, with violaceous hue arranged in a linear pattern following Blaschko's lines. The length of the lesions ranged from 5 cms to 20 cms (nearly involving the entire limb).

Sites of involvement in Children with linear lichen planus

Table - VI (n = 15)

Site	Right	Left
Upper limb	1	1
Lower limb	2	6
Trunk	1	
Head & Neck	0	
Upper limb & Lower limb	1	
Lower limb & Trunk	3	
Total	**15**	

The lesions were common over the lower extremities especially the left. One patient presented with multiple linear lesions over the trunk and extremities.

Associated Conditions

1. Insect bite allergy
2. Tinea versicolor
3. Atopic dermatitis
4. Post inflammatory hyper pigmentation

None of them showed any mucous membrane or nail involvement, and also none of the children included in this study were positive for HBsAg.

Ten out of the fifteen children were biopsied from the recent lesions. Eight specimens showed the classical features of Lichen Planus like orthokeratosis, focal hypergranulosis, saw toothed rete ridges, irregular acanthosis, basal cell degeneration, band like lympho histiocytic infiltrates hugging the epidermis and pigment incontinence.

The remaining two had the features of lichenoid dermatitis like basal cell degeneration, superficial mononuclear cell infiltrate in upper dermis, colloid bodies and pigment incontinence.

LINEAR EPIDERMAL NEVUS

In this study group, nine children presented with linear epidermal nevus. The age group ranged between 0 to 11 years, of whom six were males and three were females with a sex incidence ratio of 6:3.

Age and Sex distribution among the children with Linear epidermal nevus : Table - VII (n = 9)

Age group	Male	Female
0-2	1	0
3-5	1	0
6-8	2	1
9-12	2	2
Total	**6**	**3**

One of the child had lesion since birth. Other cases developed lesions later in life (mostly 1 year to 11 years).

All patients except one had gradual progression of lesions. 6 out of 9 cases presented with hyperpigmented, verrucous papules and plaques which coalesced to form a linear pattern and one of them had smooth, hyperpigmented papules and the remaining two had erythematous lesions.

Among these 9 children 2 were born of 2nd and 3rd degree consanguineous marriage and none of the family members of these patients had similar lesions.

Sites and involvement in children having Linear epidermal nevus

Table - VIII (n = 9)

Site	**Right**	**Left**
Upper limb	2	1
Lower limb	1	1
Trunk	1	
Head & Neck	0	
Lower limb & Trunk	1	
Upper limb & Lower limb & Trunk	1	
Upper limb & Trunk	1	
Total	**9**	

The naevi were found more commonly in the Limbs (5 cases) followed by Trunk & Limb (3 cases), followed by Trunk alone (1). Four out of nine children had nevi involving trunk implicating systematized form of verrucous epidermal nevus.

Neurological and ophthalmological evaluations were normal for all children.

Among the nine children skin biopsy was done for five cases, which showed the features of Epidermal verrucous nevus like hyperkeratosis, moderate irregular acanthosis, well formed granular layer, increased pigment basal layer, and patchy inflammatory infiltrates in upper dermis. Two of them showed the features of ILVEN like hyperkeratosis with foci of parakeratosis, moderate acanthosis, elongation and thickening of the rete ridges with a ‘posriasiform’ appearance, papillomatosis, and slight spongiosis with exocytosis of lymphocytes.

LINEAR MORPHOEA

In this study 5 cases presented with Linear Morphoea in the age group between 3 and 12 years with an average of 10.

Out of the 5 cases 2 were males and 3 were females forming a Male : Female ratio of 2:3.

Age and Sex distribution in children with Linear Morphoea

Table - IX (n = 5)

Age group	Male	Female
0-2	0	0
3-5	1	1
6-8	0	0
9-12	1	2
Total	**2**	**3**

The lesions started in early childhood or in adolescence. The duration of lesions varied from 1 year to 2 years. Four out of the five cases presented without any specific complaints and the remaining one had pain over the Morphoea. One child had prior history of intramuscular injection over the lesional site.

Two children were born of consanguineous marriage (second degree consanguinity). There was no family history of similar illness.

Two children had lesions involving the lower limbs (left side) while all the other children had lesions over the head and neck.

The lesions were skin coloured to brownish, atrophic, indurated plaques of the size of 3cms to 10cms arranged in a linear pattern

following Blaschko's lines. One child of Linear Morphoea was found to be fixed to the underlying structures. All of them had hair loss over the plaques. None of them showed either mucous membrane or nail involvement.

On investigation, Eosinophilia was seen in 2 children X-ray, ECG, EEG and Neurological opinion were sought for all the children and were found to be normal. Rheumatologist opinion was obtained for all children. Biopsy was done in 3 children of which one showed the features consistent with early Morphoea like heperkeratosis, atrophic epidermis, increased pigment basal layer eosinophilic, oedematous collagen bundles in upper dermis and cut section of eccrine duct and arrector pilorum muscle.

Two specimens showed the features consistent with Late Morphoea like atrophic epidermis, collagen bundles appeared homogenous, thickened, and hypo cellular, atrophic eccrine glands narrowed blood vessels and elastic fibers are thickened and arranged parallel to epidermis and collagen fibers.

LINEAR PSORIASIS

In this study, two children presented with hypopigmented, scaly plaques of six months duration over the lower extremity. There was no evidence of any trauma preceding the lesions. One child had scalp psoriasis and other had no similar lesion anywhere else but had similar episode 2 years back. Both had pitting over the finger nails. Skin biopsy was done for both the children, which was consistent with psoriasis like hyperkeratosis, parakeratosis, absent granular layer, regular elongation of rete ridges, with bulbous thickening at their lower ends, suprapapillary thinning of stratum malphighi, irregular dilated tortuous vessels in dermal papillae, and perivascular lymphocytes in upper dermis.

SEGMENTAL VITILIGO

In this study group two male and two female children presented with segmental vitiligo between the age group of 6 years and 12 years. Two children had lesions over the upper limb, one had over the lower limb and the other over the face.

The lesions were isolated macules to patches of size 0.5cm to 8cms. All the lesions were unilateral in distribution along the lines of Blaschko. The duration of lesions varies from 5 months to 1 year. The

lesions were asymptomatic in all children. None of them had vitiligo elsewhere or any other associated auto immune disorders.

Two children had leukotrichia over the vitiligo patches. Examination of nails and mucosa were found to be normal.

Biopsy was done for 3 children which showed the features consistent with vitiligo like absence of melanocytes in lesional skin, decreased number of melanocytes in perilesional skin and lymphocytic infiltrates in dermis.

DISCUSSION

In this study of 60 cases with a linear distribution of the lesions which did not exhibit Koebner's phenomenon, none of the cases seemed to follow the linearity determined by the Nerves, vascular or lymphatic structure and it has been suggested that these lesions follow in the lines of Blaschko.

Hence the various nevoid and acquired conditions which are supposed to follow the lines of Blaschko, which are thought to be due to a form of human mosaicism were included in this study. Most of the children were asymptomatic and mainly came for cosmetic reasons, except the Linear lichen planus children and few of Lichen striatus children who had attended our skin Out Patient Department for Moderate itching.

LICHEN STRIATUS[25,26,28]

Lichen striatus formed the majority of cases amounting to 25 in this study. The condition is said to occur commonly in the age group of 5-15 years, where as in the present study, majority of children were in the age group between 6-12 years. It was more commonly observed in Females in this study group with a Male : Female ratio of 2:3 as also been reported by Kennedy and Rogers et al[28]. According to their study there was a 2:1 female to male ratio, and the age range in this pediatric

population was 9 months to 9 years. The lesions were normally asymptomatic, with occasional pruritus in the study group as described in the literature. There were no predisposing factors in any of the children as Lichen striatus of unknown etiology[26].

Most of the children in the study group had lesions over the extremities, but few patients had lesions also over the trunk as recorded in the literature[25,26], which complied with the variable sites of expression. All the children had unilateral distribution of the lesions. None of the patients showed nail changes among this group, although changes in the form of longitudinal ridging, subungual hyperkeratosis, splitting and onycholysis have been documented[26].

Atopy was found to be associated with Lichen striatus in 80% of patients, although none in this study group had personal or family history of atopy but associated lesions like Tinea versicolor, xerosis were seen, which were not documented so far and may be coincidental.

Histopathological examination showed a chronic dermatitis picture in majority of children in this study group, few children showed psoriasiform dermatitis like features and some other showed lichenoid dermatitis like picture which was consistent with the variable histological pictures as described in the literature.

There were no systemic abnormalities noted in any of the Lichen striatus children in this study group.

LINEAR LICHEN PLANUS[74,83]

Linear lichen planus formed the next common condition in this study group consisting of 15 children. Among them most of the children were in the age group between 9-12 years and the average age at the time of diagnosis was 10 years. In this study Male : Female ratio of 8:7 was noted, showing a slight male preponderance as recorded in the literature[83].

There was no history of contact with any chemicals or trauma and 4 childrens were treated for chronic suppurative otitis media prior to the skin lesions.

There was no history of similar lesions in their family members or any other associated autoimmune disorders. It has been suggested that the linear distribution seen could be due to the tendency of Lichen planus to develop, with the formation of a clone of predisposed or vulnerable cells, which is predetermined during embryogenesis.

Most of the children reported for moderate to severe itching. The lesions started as hyperpigmented, discrete, flat topped papules with a violaceous hue in a linear pattern. In some of the children, the papules coalesced to form linear plaques which were continuous or interrupted.

In all the children there were no nail changes or mucous membrane involvement suggesting that they were all cases of isolated Linear lichen planus.

In the most of children, the lesions were found on the extremities. One child showed multiple linear lesions following the lines of Blaschko. The length of the lesions ranged from 5cms - 20cms and 1 child had lesions extending the entire length of the limb.

On histopathological examination, most of the lesions showed the classical features of Lichen planus as described in the literature[74] and one picture showed normal epidermis, basal cell degeneration, superficial mononuclear cell infiltrate in upper dermis, colloid bodies and pigment incontinence which was fit into the features of lichenoid dermatitis.

HBsAg sero positivity was not found in any one of the child which included in this study. Some of the associated features were Insect bite allergy, Tinea versicolor and Eczema which may be coincidental.

LINEAR EPIDERMAL NAEVUS[54]

In this study; Linear epidermal nevus accounted for 9 cases, with 4 cases between the age group of 9 and 12 years. Out of whom 6 were males and 3 were females. A male preponderance was seen in this study,

in contrary to equal sex incidence given into literature[54]. The children with this disorder mainly came for cosmetic reason. No family history of similar lesion was recorded.

The lesions manifested as hyperpigmented verrucous papules arranged in a linear continuous or interrupted bands and was present since birth in one of the case, but in most of the children the lesions became apparent later in childhood.

In this group 3 children had lesions involving trunk and extremities, implicating systemized form of verrucous epidermal nevus.

Most of the skin biopsies showed the classical features of verrucous epidermal nevus like hyperkeratosis, moderate irregular acanthosis, papillomatosis, well formed granular layer, increased pigment basal layer and patchy inflammatory infiltrates in upper dermis. One of them showed the features of Inflammatory Linear Verrucous Epidermal Nevus like hyperkeratosis with foci of parakeratosis, moderate acanthosis, elongation and thickening of the rete ridges with a ‘psoriasiform’ appearance, papillomatosis, and slight spongiosis with exocytosis of lymphocytes.

LINEAR MORPHOEA[64,84]

5 cases of Linear morphoea were recorded, among whom 3 were between 9-12 years of age and 2 were between 3-5 years of age. Generally the peak incidence of this condition is between 9-12 years of the age group[64]. Out of the 5 childrens, 2 were males and 3 were female children forming a Male : Female ratio of 2 : 3, correlating with the female preponderance of this condition as recorded in the literature[64].

There was no history of any provocative factors like trauma of drug intake, etc. prior to the onset of lesion except one child who had prior history of intramuscular injection over the lesional site.

There was no history of similar lesion in the family members. Most of the children presented with an asymptomatic atrophic plaques except one who had pain over the plaque.

Most of the children had lesions over the head and neck region and only 2 children had lesions over the lower limb, which is the commonest site of involvement shown in literature[84].

All the investigations pertaining to Morphoea were found to be normal including X-ray, except 2 children who had eosinophilia. Most of the skin biopsies showed the classical features of morphoea.

LINEAR PSORIASIS[48]

In this study, one child had presented with hypopigmented scaly plaque of 5 months duration over the extremity. There was no evidence of any trauma preceding the lesion. The child had fine, regular pitting over the finger nails. On histopathological examination the lesion showed a characteristic feature of psoriasis, with which, the diagnosis was revised from lichen striatus to linear psoriasis[48].

Another child presented with asymptomatic, erythematous plaques over the extremity but on histopathological examination, it was found to have hypergranulosis and orthokeratosis, alternating with absent granular layer and parakeratosis and the other characteristic features of psoriasis.

SEGMENTAL VITILIGO[88]

In this study group, two male and two female children presented with segmental vitiligo between the age group of 6 years and 12 years, which was earlier than the other types of vitiligo.

The lesions manifested as macules and patches, arranged in a linear, continuous or interrupted bands, involving mainly the extremities. One had lesions over the face and neck region corresponding to the Dermatomes rather than Blaschko’s lines, perhaps

in keeping with a neuronal etiological theory of vitiligo and it could be also a clonal susceptibility of melanocytes to neurons[88].

Two children had leukotrichia over the vitiligo patches. There was no family history of similar lesions or any mucosal or nail involvement. None of them had vitiligo elsewhere or any other associated auto immune disorders. Biopsy of these lesions showed the features consistent with vitiligo like flaky hyperkeratosis, normal epidermis and absence of melanocytes in the basal cell layer and sparse inflammatory infiltrates in upper dermis.

CONCLUSION

The following conclusions were drawn from this study.

1. The Incidence of childhood Linear Dermatoses in the skin Out Patient Department, Govt. Stanley Hospital, Stanley Medical College, Chennai during the period of Sep.2006 to August 2008, 0.66%.

2. Among the childhood Linear Dermatoses, Lichen striatus was found to be more common (41.7%).

3. The other Dermatoses following Blaschko's lines, in the descending order of frequency seen in this study were Linear Lichen Planus (25%), Linear Verrucous Epidermal Nevus (15%), Linear Morphoea (8.3%), Linear Vitiligo (6.7%), Linear Psoriasis (3.3%).

4. Incidence wise both lichen striatus and linear morphoea was found to be more in the female children, where as linear lichen planus and linear epidermal nevus showed slight male preponderance. The incidence of linear psoriasis and segmental vitiligo were found to be equal in both the sexes.

5. The importance of histopathological correlation is very obvious. Cases which were clinically diagnosed as Lichen Striatus showed histopathological features of Psoriasis and Linear Epidermal Verrucous Nevus, ultimately changing the management in any given condition. So, it is imperative to do histopathological study in all linear lesions as the line of management varies in each condition.

6. Majority of children showed unilateral distribution in a linear pattern, more often on the extremities.

7. Very few associations were noted such as cases of Lichen Planus which were associated with Insect bite allergy and Tinea versicolor and Lichen striatus with Xerosis, Tinea Versicolor.

8. The lesions were more of a cosmetic concern in most of the children in this study.

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PROFORMA

1. OP/IP No. : Serial No. :
2. Name : Date :
3. Age : Address :
4. Sex :
5. Occupation :
6. HISTORY OF PRESENT ILLNESS :
   - Complaints :
   - Duration :
   - Onset : sudden / insidious /progressive
   - Distribution :
   - Remission & exacerbation :
   - Precipitating factors : Trauma / Photosensitivity / Drug intake Soap & other cosmetics used / Infections / Stress / Others.
   - Similar skin conditions in other site :
   - Any other skin conditions:
     - Exposure to STD : Present / Absent
     - Atopy :
9. PAST HISTORY
   - History of similar illness :
   - History of any other skin disease :

10.FAMILY HISTORY

- Number of siblings :
- Consanguinous / non consanguinous :
- Other members affected :

11.PERSONAL HISTORY :

12.TREATMENT HISTORY :

13.GENERAL EXAMINATION : Anemia / Jaundice / Cyanosis / Clubbing / Pedal edema / Lymphadenopathy / Asso Congenital Anomalies.

- CVS : PR : BP :
- RS : Eye :
- CNS : Dental :
- ABDOMEN: Skeletal :

14.DERMATOLOGICAL EXAMINATION

- Basic (Primary) Morphology : Hyperpigmented/Hypopigmented Scaly / Inflammed / Atrophic / Hypertrophic Macule / Papule / Nodule / Plaques / Verrucous / Vesicle / Comedones Vascular / Hair Over the Lesion

- Secondary changes : Linear :

- Sites Affected (Topography) Upper Limb
  Lower Limb
  Trunk
  Head and Neck

- Colour : Hypopigmented
  Skin coloured
  Blue
  Black
  Brown
  Violaceous

- Involvement of Underlying structure : Yes / No
- Associated Skin Conditions :
- Associated other conditions / Malignancy :
- Whether the Lesions following
  Along the lines of BLASCHKO
  Koebner's Phenomenon
- Examination of
  Hair : Nails :
  Palms : Soles :
  Mucous Membranes :

15.INVESTIGATIONS

- Blood : Hemoglobin : GM %
  Total Count : Cells/Cu. MM
  Differential Count : N E B L M
  ESR ½HR / 1 HR :
  Platelet Count :
- Serology :
- Urine : Albumin
  Sugar
  Deposits
- Motion : OVA / CYST /
- Mantoux Text : HBsAg :
- X-ray Chest / CT Scan (If necessary) :
- Skin Biopsy No :

16.CLINICAL PHOTO :

17.PROVISIONAL DIAGNOSIS :

REFERENCE TO THE MASTER CHART

S.N.	-	Serial Number
M	-	Male
F	-	Female
Consan	-	Consanguinity
N	-	Normal
Colour code	-	1- Hypo pigmented
		2 - Skin coloured
		3 - Blue
		4 - Black
		5 - Brown
		6 - Violaceous
Distribution	-	1 - Upper Limb
		2 - Lower Limb
		3 - Trunk
		4 - Head and Neck region
Duration	-	W - Week
		M - Month
		Y - Year
P	-	Progressive
S	-	Static
Asso. skin	-	Associated skin lesions
B	-	Along the Blaschko's lines
H	-	Hair
N	-	Nail
T	-	Teeth
MM	-	Mucous membrane
LS	-	Lichen striatus
LP	-	Lichen planus
LEN	-	Linear epidermal nevus
Psora	-	Linear Psoriasis
L.Mor	-	Linear Morphoea
Ppt	-	Precipitating Factors
R	-	Routine
SP	-	Specific