CLINICAL STUDY ON PALMOPLANTAR KERATODERMA

Dissertation Submitted in

fulfillment of the university regulations for

MD DEGREE IN

DERMATOLOGY, VENEREOLOGY AND LEPROSY

(BRANCH X11 A)

THE TAMILNADU DR.M.G.R.MEDICAL UNIVERSITY

CHENNAI

APRIL 2012

CERTIFICATE

Certified that this dissertation entitled **CLINICAL STUDY ON PALMOPLANTAR KERATODERMA** is a bonafide work done by Dr.P.S.MOHANASUNDARI, Post Graduate Student of the Department of Dermatology, Venereology and Leprosy, Madras Medical College, Chennai – 600003, during the academic year 2009 – 2012. This work has not previously formed the basis for the award of any degree.

Dr.S.Jayakumar, M.D.,D.D.,
Professor and Head of the Department,
Department of Dermatology and Leprosy,
Madras Medical College,
Chennai – 600003.

Dean,
Madras Medical College,
Chennai – 600003.

DECLARATION

I, **DR. P.S.MOHANA SUNDARI,** solemnly declare that dissertation titled, "**CLINICAL STUDY ON PALMOPLANTAR KERATODERMA"** is a bonafide work done by me at the Department of Dermatology and Leprosy, Madras Medical College, Chennai-3 during the period of October 2009 to September 2011 under the supervision of **Prof. DR.S.JAYAKUMAR, M.D, D.D,** Professor and HOD, The Department of Dermatology and Leprosy, Madras Medical College, Chennai. The dissertation is submitted to Tamilnadu Dr.M.G.R. Medical University, towards partial fulfilment of requirement for the award of **M.D. Degree (Branch-XII A) in DERMATOLOGY, VENEREOLOGY AND LEPROSY**.

(**Signature of the Candidate)**

Place: Chennai

Date:

SPECIAL ACKNOWLEDGEMENT

My sincere thanks to **Prof. Dr. V. KANAGASABAI. M.D** Dean, Madras Medical College for allowing me to do this dissertation and utilize the institutional facilities.

ACKNOWLEDGEMENT

I am gratefully indebted to Prof. Dr.S.Jayakumar M.D., D.D., Professor and Head, Department of Dermatology and Leprosy for his invaluable guidance, motivation and help throughout the study. I would like to express my sincere and heartfelt gratitude to Prof.Dr.V.Sudha M.D.,D.V., Director, Institute of Venereology for her guidance.

I am grateful to Dr.S.Nirmala M.D.,D.D., Additional Professor, Department of Dermatology and Leprosy for her invaluable guidance and help. I sincerely thank DR.R. Arunadevi M.D, DD for her guidance throughout the study. I thank Dr.C. Janaki M.D., D.D. Additional Professor of Dermatology (Mycology) for her priceless support. I express my sincere gratitude to Dr.V.Sampath M.D., D.D. Dr.Priyavathani M.D., D.D., Additional Professors, Department of Dermatology and Leprosy. I also thank Dr.P.Elangovan, M.D,DV Additional professor Institute of Venerelogy for his motivation and support.

I wish to thank Dr.D.Prabhavathy M.D.,D.D., Former Professor, Department of Dermatology and Leprosy, Dr.V.Somasundaram M.D., D.D., Former Professor Department of Dermatology and Leprosy and Dr.K.Gajendran M.D., D.V., Former Director, Institute of Venereolgy for their constant support and motivation.

My heartfelt thanks to Dr.Afthab Jameela Wahab M.D., D.D., Assistant Professor, for her valuable support and guidance. My sincere thanks to Dr,J,Manjula M.D.,DNB., Dr.G.K.Tharini M.D., Dr.R.Madhu M.D.,D.C.H., Dr.S.J.Daniel M.D.,DVL., Dr.C.Vijayabhaskar M.D.,D.CH, Dr.Saravanan M.D., DVL., and Dr.S.Madhavi M.D.,DVL., Assistant Professors, Department of Dermatology for their kind support and encouragement.

I am inclined to thank Dr.V.Thirunavukarasu M.D.,D.V., Dr.P.Mohan M.D.,D.V., Dr.V.N.S.Ahamed Shariff M.D (DVL)., Dr.P.Prabhakar M.D (DVL)., Dr.K.Umamaheswari M.D (DVL)., Dr.R.Sowmiya M.D (DVL)., Dr.C.Vidya M.D (DVL)., Assistant Professors, Department of Venereology for their help and suggestions.

I thank my former Assistant Professors Dr.S.Kumaravel M.D.,D.D., Dr.A.Hamedullah M.D.,D.D., Dr.N.Hema M.D., Dr.K.Venkateswaran M.D.,D.V., Dr.S.Kalaivani M.D.,D.V., Dr.S.Arunkumar M.D., for their valuable support.

I duly acknowledge the paramedical staff and my colleagues for their help and favour. Last but not the least I am profoundly grateful to all patients for their co-operation and participation in this study.

CONTENTS

Sl.No.	Title	Page No.
1.	INTRODUCTION	1
2.	REVIEW OF LITERATURE	3
3.	AIM OF THE STUDY	47
4.	MATERIALS AND METHODS	48
5.	OBSERVATIONS AND RESULTS	52
6.	DISCUSSION	63
7.	CONCLUSION	69
8.	ANNEXURES	
	REFERENCES	
	PROFORMA	
	MASTER CHART	
	ABBREVATIONS	

INTRODUCTION

Palmoplantar keratodermas are a heterogenous group of disorders characterized by hyperkeratosis of palms and soles. They may be inherited or acquired disorders. Clinically there are three major patterns of involvement, diffuse, focal and punctate types. The differentiation between the different types depends on the presence or absence of transgrediens, erythematous borders, hyperhidrosis and systemic involvement.

Acquired PPK occur later in life, and attributable to an underlying etiology like inflammatory and reactive dermatoses, infections, drugs, systemic diseases and internal malignancy.

In the evaluation of PPK, a thorough history taking including the family history is essential to establish clinical diagnosis to assess the prognosis, to take therapeutic decisions and offer genetic counseling. The terminology and nosology of inherited PPK are continuously evolving. Advances in molecular genetics may help in further simplification.

As they are rare disorders, literature reports are based on individual family reports. Only few studies are available about the prevalence of palmoplantar keratoderma in the Indian population. Systematic analysis on the etiology of PPK's has not been done so far in our population. Hence we have undertaken this study to analyze about PPK in detail.

REVIEW OF LITERATURE

Palmoplantar keratodermas represents a diverse group of hereditary and acquired disorders in which there is hyperkeratosis of the palms and soles occurs. Keratins are the major intermediate filaments of the keratinocytes. There are two types of keratins acidic or type 1 (K9-K20, K31-K38)[3] and basic or type 2(K1-K8,K81-86)[3]. Palmoplantar epidermis is a specialized epidermis. K9 is unique to palmoplantar skin[3]. K6a and K16 are also found in nail, hair follicle, sweat gland, oral mucosa and larynx in addition to palmo plantar epidermis.

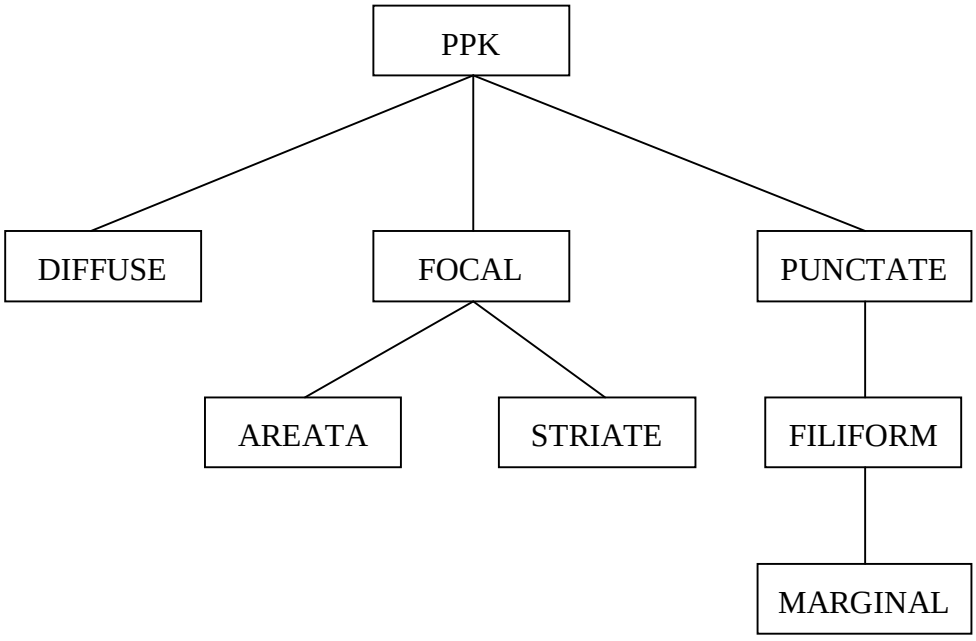
Any inherited gene defect of the structural component of the keratinocytes, keratin intermediate filaments, desmosomal proteins including desmoplakin, plakogloblin, plakophilin and desmosomal cadherin may result in keratodermas[2]. Mutations of junctional proteins and gap junctions are also implicated. Abnormality of non strucutural proteins like SLURP1-Protein, metabolic defect in Tyrosine aminotransferase and phenolic acid abnormalities also results in PPK.

The mutations occurring in these proteins lead to unstable filament network and this is responsible for hyperkeratosis of palms and soles[2]. Codistribution of keratin protein such a K6a, K16, etc. and others like connexins may explain the involvement of other systems[2].

CLASSIFICATION OF PPK

1. Simple: When found alone
2. Complex: In combination with skin lesions elsewhere
3. Syndromic: In association with complex syndrome including Ectodermal Dysplasia.

CLASSIFICATION BASED ON CLINICAL PATTERN



Diffuse : Uniform involvement of palms and soles.

Focal : Limited involvement restricted to pressure points.

1.Areata/ Nummular type (oval lesions).

2.Striated type (linear lesions).

Punctate : Small keratotic papules seen as tiny hard rounded bumps of skin on the palms and soles.

Variants:

Filiform: Tiny hard ‘spikes’ growing out of the skin of the palms and soles and Sometimes found elsewhere on the skin.

Marginal: Tiny bumps along the border of the palms and fingers, soles and toes.

CLASSIFICATION BASED ON THE EXTENT OF SPREAD

Transgrediens: Extent beyond the palms and soles (from palms onto the wrist, dorsum of hand, knuckles, elbows and from soles onto the feet).

Progrediens: Progression with age.

HISTOPATHOLOGY

The histologic changes are largely nonspecific and or confined primarily to the epidermis. Skin biopsy is done from weight bearing area, thenar and hypothenar eminence. Epidermolytic PPK (EPPK) is characterized by hyperkeratosis, acanthosis, papillomatosis. Epidermolysis at spinous, granular cell layer is noted by light microscopy. Thickened granular layer with irregularly shaped keratohyaline granules are present[4,7]. Electron microscopy shows keratin aggregates and cytolysis.[2,4.]

Nonepidermolytic keratoderma (NEPPK) is characterised by orthokeratosis, acanthosis and well preserved granular layer[4,8]. All the keratodermas are difficult to distinguish histologically and clear differential diagnosis is difficult.

CLASSIFICATION

The hereditary PPKs have been reviewed extensively in the literature and in 1996, Stevens et al proposed a classification scheme for these disorders[9]. More recently in 2005, Itin and Fistrol classified the hereditary PPKs according to inheritance pattern and molecular basis[10].

CLASSIFICATION OF HEREDITARY PPK BASED ON CLINICAL FEATURES

Type	***Disease***	***Associated features***
Diffuse	Unna-Thost - NEPPK Vorner EPPK Meleda – Mal de Meleda Greither–transgrediens & progrediens Gamborg Nielsen (Norbotten) Sybert	
Diffuse	Vohwinkel syndrome	Sensorineural deafness Knuckle pads,
	Bart-Pumphrey syndrome	Leukonychia, sensorineural deafness
	Huriez syndrome	Scleroatrophy
	Clouston syndrome	Hidrotic ectodermal dysplasia
	Olmsted syndrome	Mutilation, periorificial plaques
	Papillon-Lefevre syndrome	Periodontitis, recurrent pyogenic infections
	Haim-Munk syndrome:	Periodontitis, arachnodactyly & acro osteolysis
	Naxos disease	Woolly hair, Right Ventricular Cardio Myopathy
Focal	Striate – NEPPK, Watchers, Brunauer-Fuhs-Siemens Nummular-EPPK, Hereditary painful callosities	

Type	*Disease*	*Associated features*
Focal	Howel-Evans syndrome	Oesophageal carcinoma,
	Richner-Hanhart syndrome	Oculocutaneous tyrosinemia
	Pachyonychia congenita	
	Type1-Jadasson-Lewandowsky syndrome	Hypertrophic nail dystrophy, oral leucokeratosis and epidermal inclusion cyst.
	Type2-Jackson Lawler syndrome	Natal teeth, steatocystoma & pili torti
	Carvajal syndrome	Woollyhair& Biventricular cardiomyopathy
Punctate	Buschke-Fisher-Brauer type Acrokeratoelastoidalis	
	Punctate porokeratosis	

PPK & OTHER ASSOCIATED GENODERMATOSES

Disorders	***Syndrome***
Ichthyosis	Ichthyosis Vulgaris Bullous congenital ichthyosiform erythroderma, Non-bullous ichthyosiform erythroderma, Lamellar ichthyosis, Harlequin ichthyosis, Ichthyosis hystrix, Netherton syndrome, Sjogren Larsson syndrome, Refsum syndrome, CEDNIK (Cerebral dysgenesis, Neuropathy, Ichthyosis, Keratodermas)[3]
Ectodermal dysplasias	Ectodermal Dysplasias With skin fragility, Rapp – Hodgkins syndrome, Schopf-Sculz Passarge syndrome, Oculo dental digital dysplasia HOPP syndrome – hypotrichosis, onychrogyphosis, periodontitis & PPK Cardio facio cutaneous syndrome
Pigmentary Disorders	Naegeli - Franceschetti - Jadassohn syndrome, Dermatopathia pigmentosa reticularis, Cantu syndrome Cole disease-guttate hypopigmentation with punctate PPK.
Mechano bullous disorders	Epidermolysis bullosa simplex, Dowling Meara with mottled pigmentation, Weber-Cockayne, Non Herlitz junctional type, Kindler syndrome
Epidermal nevus associated with linear PPK	
Disorders of keratinisation	* Erythrokeratodermas a) Erythrokeratodermas variabilis[3] b) Progressive symmetric Erythrokeratodermas * Familial pityriasis rubra pilaris * Dyskeratosis congenita

PPK AND THEIR GENETIC BASES

Types	*Inheritance*	*Gene defect*
Vorner syndrome	A.D	Keratin 1 & Keratin 9
Unna – Thost	A.D	Keratin1,Chromosome12q11-q13
Loricrin keratoderma	A.D	Loricrin, Connexin26, chromosome 1q
Greither's disease	A.D	Keratin 1
Sybert's Keratoderma	A.D	Keratin 1
Bart Pumphrey syndrome	A.D	Connexin 26
Huriez syndrome	A.D	chromosome 4q23
Clouston syndrome	A.D	Connexin 30-GJB6
Olmsted syndrome	A.D. & X.L.R.	Abnormal expression of K5 &K14
Mal de Meleda	A.R	SLURP -1(secreted Ly-6/uUPAR related protein)
Papillon-Lefevre syndrome	A.R	Cathepsin-C,chromosome 11q14.1-q14.3
Haim-Munk syndrome	A.R	Cathepsin C
Naxos disease	A.R	Plakoglobin
Focal keratoderma Striate /Areata type	A.D	Desmoglein1, desmoplakin K1 chromosome 18q12.1

Types	Inheritance	Gene defect
Pachyonychia congenita		
Type 1: Jadassohn-Lewandowsky	A.D	Keratin 6a &16
Type 2: Jackson-Lawler	A.R	Keratin 6b &17
Carvajal syndrome	A.R	Desmoplakins
Schöpf-Schulz-Passarge syndrome	A.D	Locus not identified
Punctate type:		
Buschke-Fisher-Brauer syndrome	A.D	Chromosome 15q22.2 & 15q22.31

DIFFUSE KERATODERMAS-NON TRANSGREDIENS TYPE

VORNER SYNDROME –EPIDERMOLYTIC PPK

This is the most common type of hereditary PPK described by Vorner and Klause et al. The incidence is about 4.4/100000 in Northern Ireland[1,3,13].

Clinical Features: The disease starts from infancy as diffuse keratodermas with sharp border, erythematous edges and waxy or verrucous surface. History of recurrent blistering or fissuring of the palms is present. Hair, teeth and nail are normal. Secondary dermatophyte infection and pitted keratolysis are common.

Associations includes raised serum IgE levels[107], knucklepads[108], nail changes[109], periodontis[110], clubbing and adenocarcinoma of the colon[111].

Histopathology: Epidermolytic hyperkeratosis is present with occasinal blister formation is seen[1,2,3].

UNNA- THOST SYNDROME -NON EPIDERMOLYTIC PPK[1,2,14.]

It was described by Thost in 1880 as a form of palmoplantar ichthyosis. Unna classified it as a variant of keratoderma in 1883. In 1962, in his monograph on acrokeratoses, Osvaldo Costa considered it to be the main representative of all types of palmoplantar keratosis.

Clinical Features: This disease is seen in all races with onset at 2-5 years of age. It is a nontransgrediens type of PPK, initially appears over soles then spreads to the palms with a yellow waxy or verrucous surface and livid red margins. There is no pruritus and it worsens in cold weather. Hyperhidrosis, secondary dermatophyte infection and pitted keratolysis are common[1]. Nails show thickening without dystrophy. Hair and teeth are normal.

Histopathology:There are non specific changes with hyperkertosis, acanthosis and reduction in thickness of granular layer. Biopsy is needed to exclude EPPK[14].

TRANSGREDIENT KERATODERMAS

LORICRIN KERATODERMA

Synonyms - Camisa Syndrome, Variant Vohwinkel's syndrome, Mutilating keratoderma with ichthyosis.Keratoderma hereditaria mutilans Vohwinkel first described this disorder in 1929.[27]

Clinical Features: A rugose keratoderma develops during childhood, gradually becoming honeycombed pattern. Initially it is diffuse then becomes transgredient, followed by development of pseudo ainhum formation leading to auto amputation of digits. Characteritic starfish hyperkeratosis appear over the knuckles .

Generalized mild ichthyosis is present[50]. There may be a history of collodion baby at birth and desquamation is also reported. Mutations in CX26 (GJB2) is associated with profound deafness[1,3,51].

Histopathology: Hyperkeratosis, acanthosis, thickened granular layer with retained nuclei in stratum corneum is seen. Immune electron

microscopy shows presence of loricrin in these nuclei[1]. Lysosomal B glucuronidase is elevated.[112]

BART PUMPHREY SYNDROME

Bart a dermatologist and Pumphrey an otorhinolaryngologist described this entity in 1967[29].

Clinical features: Profound hearing impairement is present from birth. During early childhood diffuse PPK develops with knuckle pads over the metacarpophalangeal, proximal and distal interphalangeal joint. There is variable leukonychia, which improves with age. This syndrome can overlap with Vohwinkel syndrome due to connexin 26 mutation.[30]

HURIEZ SYNDROME

Synoyms – keratoderma with scleroatrophy.

It is a rare disorder. Less than dozen families have been described.

Clinical Features: Palms are more affected than soles. Red atrophic skin over dorsum of hands and feet is present. Pseudo sclerodermatous change with scleroatrophy is seen. There is increased risk of developing squamous cell carcinomas over atrophic areas.[32] Nail findings include ridging, koilonychica, fissuring and hypoplasia.

Histopathology: Biopsy shows hyperkeratosis, acanthosis, increased granular layer and complete absence of Langerhans cells[1,3]. On electron microscopy, dense bundles of tonofilaments and clumping of keratohyaline granules are seen.

MAL DE MELEDA[35,36,37]

Synonyms- Keratoderma palmoplantaris transgrediens, Acroerythrokeratoderma.

It is named after the Croatian island of Meleda(Mljet) where it was first identified. The incidence in the general population is 1 in 10,000.[34]

Clinical features: Onset of this disease is shortly after birth. Diffuse progressive palmoplantar thickening of transgrediens type with an erythematous border is present. Waxy yellow hyperkeratotic lesions over elbows and knees are seen. Severe hyperhidrosis accompanied by malodor is characteristic of this PPK[35]. It is complicated by secondary bacterial and fungal infection.

Periorificial lesion resembling Olmsted syndrome is seen. Nail shows thickening, kolinonychia and subungual hyperkeratosis.

Sclerodactly and constricing bands around the digits may occur. Angular chelitis is common. Melanoma within the affected skin is reported[37].

Histopathology: There is thickening of stratum corneum, stratum lucidum with marked acanthosis, normal granular layer, enlarged Sweat glands and prominent perivascular lympho histiocytic infiltrates.

GREITHER'S DISEASE (transgrediens et progrediens)

Greither first described this disorder in 1952.

Clinical features: Onset is in early infancy with diffuse PPK showing an erythematous border. The lesions tend to extend to the dorsum of the hands and feet (transgradiens), and hyperkeratotic plaques are present on the elbows and knees. Marked hyperhidrosis is said to be typical of this condition. This disease may improve with age. Greither described a case with gradual onset which improved by 5^{th} decade[1].

Histopathology: The histology described by Greither and others is not specific. An electron microscopic study by showed aggregated tonofilaments around the nucleus, without true clump formation[34]. Desmosomes were numerous and cell-cell junctions showed an imbricated pattern.

SYBERT 'S KERATODERMA

This was reported by Sybert et al as a more severe trangredient keratoderma[26].

Clinical features: This is Similar to Greither's keratoderma with an earlier onset. Glove and stocking hyperkeratosis extend to the elbows, knees and natal cleft. Auto amputation of toes may occur[24].

Histopathology: Excessive lipid laden cells are seen in stratum corneum. Electron microscopy shows abnormal distribution of keratohyaline granules.

CLOUSTON SYNDROME (HIDROTIC ECTODERMAL DYSPLASIA)

Clouston described this as a distinct type of ectodermal dysplasia It is more common among people with French- Canadian ancestry[46].

Clinical feature: PPK initially develops over pressure points and increases in extent with age. Hyperkeratotic plaques appear over elbows, knees and knuckles. Localized hyperpigmentation is a striking finding over the above areas. Skin is dry and rough. Sweating is normal. Oral leucoplakia is observed.

There is diffuse hypotrichosis involving the scalp hair, eyebrows, eyelashes, axilla and genital region. Total alopecia can occur. Ultra structural examination of hair shows disorganization of hair fibres and loss of cuticles.[1]

Nails are normal at birth become thickened and dystrophic and are easily shed. It mimics Pachyonychia Congenita[47]. Teeth are normal but caries is common. Various eye abnormalities such as strabismus and congenital cataract and sensory neural hearing loss have been reported.

Histopathology: Histology of this PPK is nonspecific. In hairy skin, the hair follicles are absent, reduced or dystrophic. Sebaceous and apocrine glands are sparse or absent. Eccrine glands are normal[1].

OLMSTED SYNDROME (CONGENITAL PALMOPLANTAR AND PERIORAL KERATODERMA)

This is a very rare disorder, named after H. C. Olmsted. 50 cases have been described so far in literature[48].

Clinical features: This disease begins during infancy. During the first year well defined erythematous hyperkeratotic plaques appear around perioral,[52] inguinal, genital and gluteal cleft. There is a combination of bilateral, mutilating, palmoplantar keratoderma with

flexion deformities of the digits, leading to constriction or spontaneous amputation. Linear keratosis is observed over the forearm. Alopecia, nail changes, tooth anomaly, joint laxity and corneal dystrophy are also seen.

Presence of Palmoplantar keratoderma along with perioral and perinasal keratotic plaques excludes other syndromes of keratodermas including Mal de Meleda, Vohwinkel syndrome and Pachyonychia congenita. Failure to respond to oral Zinc therapy rules out the possibility of acrodermatitis enteropathica.

The disease has a slow but progressive course. Squamous cell carcinoma and melanomas are observed in areas of keratodermas[49]. The periorificial lesions may or may not improve with age.

PAPILLON-LEFEVRE SYNDROME

It was first described in 1924 by two French physicians, Papillon and Lefevre[41].

Clinical Features: This is characterized by palmoplantar hyperkeratosis, periodontopathy and precocious loss of dentition. Onset of PPK is around 1 - 4 years[40]. The sharply demarcated, erythematous keratotic plaques involve the entire surface of the palms and soles,

sometimes extending onto the dorsum of the hands and feet. There may be associated hyperhidrosis, which may cause a foul smell. The symptoms may worsen in winter and be associated with painful fissures. The keratosis of the plantar surface extends to the edges of the soles and occasionally onto the skin overlying the Achilles tendon and the lateral malleoli. The psoriasiform plaques may also be seen on the elbows and knees. Other sites that may be affected include the eyelids, cheeks, labial commissures, legs, thighs and axillae. The hair is usually normal but the nails, in advanced cases, may show transverse grooving and fissuring.

The second major feature is periodontitis starts at the age of 3 or 4 years[42]. There is normal development and eruption of the deciduous teeth. It is associated with severe gingival inflammation in the absence of any local etiologic factor which is unresponsive to traditional treatment modalities. Rest of the oral mucousa membrane completely normal. There is premature exfoliation of primary dentition by the age of 4 years followed by healthy gingiva. The whole process gets repeated with eruption of permanent dentition, and there is loss of

Permanent teeth by the age of 13-16 years. Later, the third molars also undergo the same fate. Severe resorption of alveolar bone gives the teeth a 'floating-in-air' appearance on dental radiographs[42].

In addition to the dermatological and oral findings, patients may have decreased neutrophil, lymphocyte or monocyte functions and an increased susceptibility to bacterial infection, leading to recurrent pyogenic infections of the skin. Pyogenic liver abscess is a complication and is associated with impairment of the immune system. Another feature is the presence of intracranial calcification in the choroid plexus and tentorium.

Histopathology:Biopsy shows hyperkeratosis, focal parakeratosis acanthosis, psoriasiform hyperplasia, tortuous capillaries in dermal papillae and superficial lymphocytic infiltration.

HAIM-MUNK SYNDROME

This is a variant of Papillon- Lefevre syndrome[1]

Clinical features: Similar findings with identical PPK and periodontosis is present in addition there is onychogryphosis,

arachnodactly, acro osteolysis and atrophic changes. Hypotrichosis is also reported. Good dental care saves the teeth in this condition[114].

Naxos Disease

It was first reported in 1986 by Protonotorios et al in patients originating from the Hellenic island of Naxos. The prevalence of the disease in Greek islands is 1:1000. It is characterized by arrythmogeneic right ventricular cardiomyopathy, NEPPK, and woolly hair.[55]

Clinical features: Onset is during the first year of life. PPK is diffuse, nontransgredient with erythematous borders. Woolly hair is present at birth. Cardiac manifestations start late after puberty in the form of arrhythmias, cardiac failure and sudden death. Cardiac histology shows loss of myocardium replaced by fibrous tissue. Whenever woolly hair is associated with PPK, search for cardiac abnormalities is to be done[54].

CARVAJAL SYNDROME

It has been described in three families from Ecquador.

Clinical features: This condition is characterized by striate PPK, woolly hair and left ventricular dilated cardiomyopathy, sometimes

biventricular cardiomyopathy. Woolly hair is present at birth. Keratoderma appears early in infancy. In addition lichenoid keratosis of the flexures, elbows and knees are present. Transient vesicles and blisters are also seen[56].

Histopathology: Widening of intercellular spaces, clustering of desmosomes and collapsed intermediate filament network are seen.

FOCAL KERATODERMAS (ISOLATED)

Synonyms –PPK striata/ areata, Focal NEPPK, Watchers PPK, Brunauer-Fuhs-Siemens PPK & Hereditary painful callosities (focal EPPK).

Clinical feature: They occur as localized circular or linear lesions on the pressure points over the palms and soles. Stress on the tissue and environmental factors aggravate the condition. Sometimes blisters may be seen. Some of these families also displayed oral leukokeratosis and follicular keratosis. Focal keratosis may be the forme fruste of Pachyonychia congenita[3]. They may be associated with other cutaneous or ectodermal disorders, including Dowling Meara epidermolysis bullosa simplex, or with disease in other systems[3].

HEREDITARY PAINFUL CALLOSITIES

They are nummular type of EPPK. Here the changes are localized to pressure points on the plantar surface and are inevitably painful. Histology shows epidermolytic hyperkeratosis[57]. Molecular abnormalitiy is not yet reported[3].

Focal Type with Associated Features

Howel – Evans Syndrome (Tylosis - Oseophageal Carcinoma)

This disorder was described by Howel–Evans[59]. It is associated with late development of oseophageal cancer. Two types are seen[3].

Clinical Features : Type- A- PPK developes by 10 years of age, often limited to pressure areas of balls of feet with mild involvement of palms. Manual labor aggravates the problem. Keratosis pilaris, oral leukokeratosis, and dry skin is often present. The risk of developing oseophageal carcinoma is higher and mostly during the 5th decade.

Type-B- Onset of PPK is at an earlier age. It carries a relatively benign course. Some people never develop oesophageal carcinoma[3].

RICHNER - HANHART SYNDROME

Synonyms - Tyrosinemia type II (Oculocutaneous Tyrosinemia)

Richner in 1938 and Hanhart in 1947 described this clinical syndrome independently[61]. Ocular lesions, painful palmo plantar hyperkeratosis and mental retardation are the cardinal features of this disease.

Clinical feature: Eye symptoms develop as early as 2 weeks of age and include redness, lacrimation and photophobia. The eye signs are corneal clouding with corneal opacities, dendritic ulcers and corneal scarring. Deposition of tyrosine crystals in the cornea is responsible for these ocular changes.

Skin manifestations usually begin after the first year of life, but may occur as early as the first month of life. The skin manifestations are well demarcated, progressive, painful, nonpruritic hyperkeratotic papules and plaques involving the soles and palms associated with hyperhidrosis. The pain in the soles may be severe enough to prevent ambulation. Mental retardation occurs in less than 50% of patients.[60]

Histopathology: skin biopsy shows hyperkeratosis ,acanthosis and hypergranulosis. ultra sructuraly keratinocyte contain clumped

tonofilaments with adeherent keratohyaline granules resembling 'dew drops on blade of grass'[1].

Investigations reveal high urinary tyrosine levels with associated high plasma tyrosine levels estimated by tandem mass spectrometric assay. The threshold levels of tyrosine for appearance of clinical manifestation is reported as 1000 umol/L. It is reasonable to keep the blood tyrosine level at 600 umol/L. Good dietary restriction of phenylalanine and tyrosine, may prevent or delay the cognitive impairment with complete resolution of skin and eye lesions.

PACHYONYCHIA CONGENITA

This disorder is characterized by dystrophic, thickened nails and focal palmoplantar keratoderma. Muller made one of the first documented observations of pachyonychia congenita in 1904.The next reports were published in 1905 by Wilson and in 1906 by Jadassohn and Lewandowsky[62].

Various classifications for pachyonychia congenita have been proposed. Currently, 2 main subtypes of pachyonychia congenita are recognized:

1) Pachyonychia congenita type 1- Jadassohn-Lewandowsky,

2) Pachyonychia congenita type 2 - Jackson-Lawler,

TYPE 1 - JADASSOHN-LEWANDOWSKI SYNDROME

This is the more common variant of pachyonychia congenita.

Clinical feature: Hypertrophic nail dystrophy (pachyonychia) is the characteristic feature of pachyonychia congenita. Although all 20 nails are usually affected, changes are more severe in the toenails, thumb and index fingers. Thickening and discoloration are present at birth or develop within the first few months of life. They may be accompanied by painful paronychia. Wedge shaped thickening due to subungual hyperkeratosis gives an omega shape to nails[62].

Symmetrical focal palmoplantar keratoderma occurs in 91-96% of patients. It usually develops in early childhood, after the nail changes, when the child starts walking and bearing weight. The keratoderma is generally worse on plantar surfaces than on palmar surfaces. Fissuring and secondary infection may also occur. Palmoplantar hyperhidrosis and follicular keratosis are frequently observed. Typical epidermal

inclusion cysts may occur. They often develop in intertriginous areas and may be numerous enough to mimic hidradenitis suppurativa.[62]

Oral leukokeratosis (benign) may be one of the earliest sign. The leukokeratotic plaques occur most commonly on the tongue and buccal surfaces of the mouth.Laryngeal involvement and Respiratory stridor has been reported. Laryngoscopic evaluation has revealed laryngeal changes that range from thickening to exophytic masses[3].

Histopathology: Biopsy findings are NEPPK with hyperkeratosis and patchy hypergranulosis. Electron microscopy shows large misformed keratohyalin granules appearing as perinuclear keratin aggregates.

TYPE 2 - JACKSON-LAWLER SYNDROME

This variant was first described by Murray. Features are similar to type 1 but the palmoplantar keratoderma may be milder and oral leukokeratoses less frequently seen. The distinguishing features are natal teeth, steatocystomas, and pilitorti.

Natal or prenatal teeth are present at birth or appear within the first 30 days of life. They are typically lost in infancy and replaced with

normal permanent teeth during childhood. Numerous steatocystomas, and a variety of cysts, including epidermal inclusion cysts, pilosebaceous cysts, and vellus hair cysts, may be seen. Pili torti or twisted hair, has been reported as a rare occurrence.[1,3]

TYPE 3 -SCHAFER –BRANAUER SYNDROME

This syndrome has similar feature of type 1 along with corneal dyskeratosis.

TYPE4 - PACHYONYCHIA CONGENITA TARDA

Similar to type 1 but has late onset in second or third decade.[64,65]

SCHÖPF-SCHULZ-PASSARGE SYNDROME

It is a very rare syndrome. So far 23 cases has been described so far in literature.[67]

Clinical features: This syndrome was first described in two sisters showing the unusual combination of multiple eyelid apocrine hidrocystomas, palmoplantar keratoderma, hypotrichosis, nail dystrophy, hypodontia and facial telangiectases. (tricho-odonto-onycho-dermal dysplasia). Multiple skin adnexal tumours are seen[1].

PUNCTATE TYPE

Clinically, it is very difficult to differentiate punctate PPK from focal types of keratoderma. The various terms used for morphological patterns of punctate Keratoderma are keratoma dissipatum, keratoderma punctata/papulosa, disseminated clavus & papulotranslucent acrokeratoderma.

Punctate PPK (keratosis punctata palmaris et plantaris)

Synonyms – Buschke-Fisher-Brauer Syndrome

Rare type with an incidence of 1.7/1,00,000. It was first described by Buschke and Fisher in the year 1910. Three years later Brauer confirmed its hereditary nature[69].

Clinical feature: The disease starts during adolescence, begins as pinpoint firm translucent papules, later becomes opaque and verrucous over time. They can enlarge to form callus like lesions. Sometimes there is a central core which on removal leaves a central depression. They are aggravated by manual work. There is no hyperhidrosis.

Associations- Anodontia, spastic paraplegia, HLA B27 associated arthropathy and internal malignancy[69].

Histopathology- Biopsy of the lesional skin shows massive hyperkeratosis over a limited area of epidermis, increased thickness of the granular layer and dermis devoid of inflammation.

PUNCTATE KERATOSIS OF THE PALMAR CREASES

Autosomal dominant type seen in African populations. Warty lesions over the palmar creases and fingers with a clavus like lesion over the medial border of distal palmar crease. Can coexist with nummular or punctate PPK.

FILIFORM TYPE (MUSIC BOX SPINE KERATODERMAS)[1]

This a rare type with autosomal dominant inheritance. The age of onset is after 20 years of life. Fine keratotic lesions project a millimeter from the palmoplantar surface. Rarely it may arise from other areas in the skin also.

Associations: Polycystic kidneys and liver[70], Darier's disease,[3] Epidermodysplasia verruciformis, Multiple myeloma, Tuberculosis, Renal failure and HIV associated PRP.

Histpathology: shows non specific change.

MARGINAL TYPE

Acrokeratoelastoidiosis of Costa and focal acral hyperkeratosis.[74] Both disease are characterized by autosomal dominant inheritance with skin coloured papules occurring on the marginal borders of the digits, hands and feet. Some papules may be umblicated. Costa described degeneration of elastic fibers. Dowd et al described the onset before the age of 20 and female preponderance.

Histopathology: Skin biopsy differentiates the two condition. Only hyperkeratosis is seen in focal acral hyperkeratosis, while degeneration of elastic fibers is noted in Costa type.

MISCELLANEOUS KERATODERMA SYNDROMES

BUREAU-BARRIRER SYNDROME

Characterized by diffuse keratoderma with osteolysis in forefoot area, polyneuropathy, painless ulcers of the feet and finger clubbing[1].

PAPILLOMATOUS VERRUCOUS PPK

Reported by Jacak and Wolf in four sibilings with autosomal recessive inheritance. The eatures include florid warty keratoderma, dysplastic teeth and follicular keratoses[1].

SYMMETRICAL INTERDIGITAL KERATODERMA

Sporadic condition of hands in which thickening of interdigital spaces occurs from 2nd decade without any occupational history and showing poor response to steroids and keratolytics[1].

CIRCUMSCRIBED HYPOKERATOSIS OF THE PALMS AND SOLES

Occurs in middle aged women. There are atrophic lesions with reduced stratum corneum, granular layer and intraepidermal vacuolation.[1]

KERATODERMA WITH NEUROPATHY

Various reports associates keratoderma with spastic paralysis. This includes striate keratoderma with pes cavus, spastic paraplegia and mental retardation. Focal keratodermas are seen in patients with motor and sensory neuropathy and nail dystrophy and Charcot - Marie-Tooth disease. Punctate keratodermas is also reported with spastic paralysis.[74]

ACQUIRED PPK

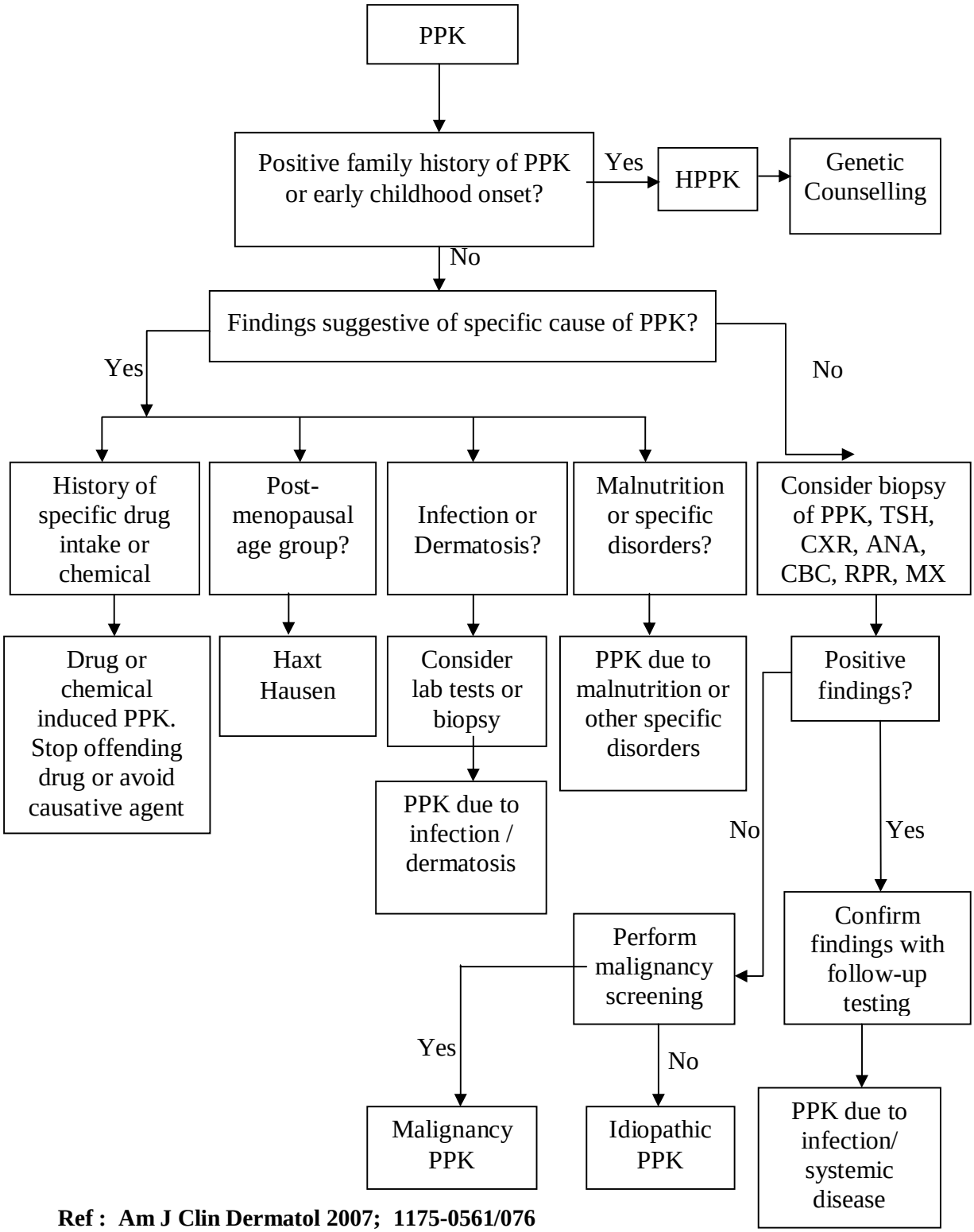
Acquired PPK is defined as a non-hereditary, non-frictional hyperkeratosis of the palms and or soles that involves more than 50% of the surface of acral areas and that may or may not be associated with clinical and histologic inflammation[6].

Acquired PPKs occur later in life, without a positive family history, and tend to be attributable to an underlying etiology. They have varying epidermal involvement patterns: The clinical pattern may be diffuse, focal, and punctate. In general, acquired keratodermas are histologically nonspecific, with marked hyperkeratosis, parakeratosis, acanthosis, hyperplasia of the stratum spinosum and granular layer, and perivascular infiltrate of chronic inflammatory cells.

ACQUIRED CAUSES OF PALMOPLANTAR KERATODERMA

Classification	Etiology
Infections & infestations	Human papilloma virus, Syphilis, Leprosy, Tuberculosis Dermatophytosis & Crusted scabies.
Dermatoses	Psoriasis, Keratoderma blennorrhagica, Pityriasis rubra pilaris, Atopic dermatitis, Chronic hand dermatitis, Lichen planus, Lichen nitidus & Lupus erythematosis.
Chemicals	Arsenic, Chloracnegens
Drugs	Glucan, Tegafur,5 fluorouracil, Bleomycin, Hydroxyurea, Lithium, Verapamil, Venlafaxine, Quinacrine (mepacrine)
Systemic diseases	Hypothyroidism, Myxedema, Chronic lymphedema, Circulatory disorders, Malnutrition
Post menopause	Keratoderma climactericum
Malignancy Acrokeratosis paraneoplastica Tripe palms	Mycosis fungoides, Sezary syndrome Esophageal cancer, Lung cancer, Breast cancer, Bladder cancer, Stomach cancer, Colon cancer, Myeloma
Aquagenic	
Idiopathic	

APPROACH TO ACQUIRED PPK



Ref : Am J Clin Dermatol 2007; 1175-0561/076

PPK CAUSED BY INFECTIOUS AGENTS

Human papilloma virus infection, especially in the immune compromised host, can appear as confluent verrucous masses on the palms and sole that mimic PPK. In syphilis, secondary lesions classically involve the palms and soles and may manifest as either diffuse or focal hyperkeratosis[91].

Leprosy, because it affects peripheral nerves, can cause hand and feet contracture and may manifest as hyperkeratotic lesions and diffuse palmoplantar thickening. Tuberculosis, especially miliary tuberculosis, has been described as presenting with palmar and plantar lesions with notable hyperkeratosis[6].

Dermatophytosis may also develop into prominent hyperkeratosis, primarily affecting the palms and soles. In crusted scabies, lesions may begin as ordinary scabies but progress to become markedly hyperkeratotic and crusted on palmar surfaces.

PPK DUE TO DERMATOSES

In psoriasis, hyperkeratosis can be both diffuse and central. Findings such as nail pitting, depressed plaques on the sides of the

fingers, involvement of the knuckles, and presence of typical plaques, together with biopsy results, may aid in making the correct diagnosis[92]. Reiter's disease can present as keratoderma blennorrhagica in some cases, which appears as compact heaped-up lesions that resemble nail heads[1,2,3,6]. Pityriasis rubra pilaris can present as thickening of the palms and soles with even, yellow hyperkeratosis, often described as having a 'carnuba wax' or PRP sandal[1,2,3,6].

Chronic hand dermatitis can be difficult to distinguish from isolated PPK. However, a history of exacerbating factors (i.e. work) and the presence of pruritus may suggest an eczematous etiology of PPK.

Lichen planus may present as a typical keratoderma with erythema and scaling or as punctate keratoses with a warty texture and yellow hue, while lichen nitidus has presented with nail dystrophy and palmoplantar hyperkeratosis in one case[91]. Chronic cutaneous lupus erythematosus may present predominantly with palmar lesions and may be dry and atrophic or hypertrophic type.

CHEMICALS

Sass et al. described two cases of arsenic induced PPK. Arsenical exposure results in punctate PPK, diffuse or spotted hypo and or

hyperpigmentation, particularly of the abdomen. Anemia and leucopaenia is present. Diagnosis confirmed by forensic examination of the hair and nail. Acitretin along with keratolytics are found to be useful[81].

Poskitt et al. described PPK in chloracegens exposure. Chloracnegens, which are halogenated aromatic compounds found in polyhalogenated naphthalenes, polyhalogenated bisphenyls, and contaminants of polychlorophenol compounds, especially herbicides. Resolution occurs 2-4 months following discontinuation of offending agent[82].

DRUGS

Drug induced PPK should be considered when PPK develops following the intake of new medications .However, the diagnosis can be clinched only when the findings resolve following discontinuation of the suspected medication.

SYSTEMIC DISEASE &PPK

Hypothyroidism is a rare cause of acquired PPK. Myxedema associated with PPK was first reported by Shaw et al. Distinctive

features of the keratoderma in these cases include, marked severity, sometimes verrucous in nature, with diffuse plantar involvement and limited involvement of the palms. However, there was a striking lack of response to topical corticosteroids and keratolytics, together with a rapid response to thyroid hormone replacement[97,98].

In chronic lymphedema, due to filariasis or lymphedema of any etiology the skin becomes diffusely thickened and develops a velvety papillomatous surface. This improve after treatment with etretinate. Other circulatory disorders, including acrocyanosis and livedo reticularis, have been also been reported in association with keratoderma[6].

KERATODERMA CLIMACTERICUM

SYNOYMS - HAXTHAUSEN DISEASE

This condition was first described by Haxthausen in 1934 in women of postmenopausal age[75]. It has been proposed to be due to hormonal dysregulation

CLINICAL FEATURES

This entity appears in women of menopausal age with no personal or family history of skin disease. A strong association with marked

obesity and hypertension has been noted. Lesions are initially round or oval in shape and of varying size and of non trangrediens type. They develop first on the plantar pressure points, with erythematous painful plaques and fissures. The condition slowly extends, becomes confluent, but is rarely associated with pruritus. Later the hyperkeratosis starts in the thenar and hypothenar eminence and becomes confluent. There is no specific response to topical oestrogen or hormone replacement therapy[76,78].

Histopathology shows compact orthokeratotic hyperkeratosis together with hypergranulosis and decreased collagen content. Diagnosis is mainly clinical. It is essential to rule out dermatophytosis and contact dermatitis. Biopsy helps in ruling out psoriasis.

PPK AND MALIGNANCY

Isolated PPK has been noted as a marker of internal malignancy .It has been described as a component of Bazex syndrome and Sezary syndrome. Cobblestone pattern with disruption of the dermatoglyphics is the common presentation.

Acrokeratosis paraneoplastica, is also known as Bazex syndrome[0]. Bazex and Griffiths first described the syndrome, which affects men

above 40 years of age and is mainly associated with neoplasia of the upper aero digestive tract[99].It is also reported in European literature in patients with prostate carcinoma, primary squamous cell carcinoma of the leg, adenocarcinoma of the stomach, and squamous cell carcinoma of the vulvar region[6].

Three stages of the disorder have been described. The eruption starts as erythema and psoriasiform scaling on the fingers and toes, then spreads to the helices and nasal bridge, leading to violaceous erythema and pityriasiform scaling. Next, it spreads to the palms and soles, producing keratoderma with a violaceous color, and giving the appearance of the honeycomb pattern. Finally if the neoplasm has not been treated, the rash extends to the legs, knees, thighs, arms, and scalp[100].

Histology is nonspecific. Symptomatic treatment occasionally results in improvement of the skin lesions, but removal of the tumor is the definitive treatment. The rash may reappear if the tumor recurs.

Tripe palms was a term first introduced by Clarke and later popularized by Breathnach and Well to denote a cutaneous paraneoplastic syndrome. The term refers to a distinctive form of palmar

keratoderma with a thickened, moss-like or velvety texture characterized by exaggerated dermatoglyphics of the palmar surface of the hands and fingers[102]. Tripe palms have been reported to occur with or without acanthosis nigricans. An underlying malignancy is found in over 90%of cases.

AQUAGENIC PPK

Aquagenic PPK, first described by English and McCollough in 1996, is a keratoderma characterized by burning and edema limited to the hands and feet after brief immersion in water[94]. It was first reported in two sisters and was termed transient reactive papulo translucent acrokeratoderma because it resembled hereditary papulotranslucent acrokeratoderma. The keratoderma is transient, recurrent, bilateral and symmetrical showing mildly translucent to whitish papules with central puncta. The eruption disappears spontaneously within minutes to hours after drying of the palms. The condition has also been associated with cystic fibrosis, and has been described in patients with the delta F508 mutation[1]. As such, cystic fibrosis should be considered in patients with aquagenic keratoderma. Avoiding water exposure in acral surfaces,

aluminum chloride hexahydrate, and barrier agents containing petrolatum have been found useful[1].

IDIOPATHIC ACQUIRED PPK

It should be reserved as a diagnosis of exclusion when all acquired etiologies have been investigated and ruled out.

CLINICAL FEATURES

They may present in any clinical pattern. It remains unknown whether the keratoderma was idiopathic or a premalignant marker. Palmoplantar filiform parakeratotic hyperkeratosis is also associated with digestive adenocarcinoma. Therefore, the diagnosis of idiopathic acquired PPK is established after through investigations.

COMPLICATIONS OF PPK

Pain due to lesions on weight bearing area, and feel of walking on pebble like mass, makes walking difficult. Fine movements of hands and fingers may be affected due to blunt sensation and the stiffness of the hands limits the occupational availability. Constricting bands,

pseudoainhum, autoamputation of digits result in further problems in daily activities[2].

Hyperhidrosis and malodour further stigmatizes and make them prone for secondary infections like tinea pedis, tinea. mannum, pitted keratolysis, and maceration of skin. They are prone to develop recurrent pyodermas..[2]

Progressive disability of other organs when associated with systemic involvement like cardiac dysrhythmias, congestive cardiac failure, deafness, ocular and dental problems results in further impairement. Poor quality of life may be due to the above factors.

TREATMENT

All types of hereditary and non hereditary keratodermas is difficult. The most common therapeutic options only result in short-term improvement and are frequently compounded by unacceptable adverse effects.

Treatment options include Supportive therapy in the form of Physical debridement, topical keratolytics, topical Retinoids, treatment

of hyperhidrosis, treatment of superficial fungal infections with topical and systemic antifungal drugs, antibiotics for secondary bacterial infections and Systemic retinoids.

Reconstructive surgery with total excision of the hyperkeratotic skin followed by grafting can also be done. Dermabrasion and CO_2 laser may be benefical in limited Keratoermas.

As the genetic basis for most of the inhereited keratodermas are known,gene therapy is possible.The knowledge about the genetic basis permit the development of prenatal diagnosis in severe or life threatening cases.

In acquired PPK, correction of underlying etiology will bring resolution of the keratoderma.Paraneoplastic keratodermas are generally refractory to local treatment and may only respond to removal of the underlying neoplasm.

AIM OF STUDY

- To study the prevalence of PPK among patients attending the Dermatology OPD, Rajiv Gandhi Government General Hospital, Chennai
- To study the etiology of PPK whether inherited or acquired
- To analyse the age distribution
- To Analyse the sex distribution
- To analyse various clinical presentations and associated symptoms

MATERIALS AND METHODS

STUDY DESIGN : Descriptive study

STUDY PERIOD: Two years from October 2009-September 2011

STUDY CENTRE: Department of Dermatology and Leprosy, Rajiv Gandhi Government General hospital, Madras Medical College, Chennai

STUDY POPULATION:

Patients with Palmoplantar keratoderma attending the Out Patient Department of Dermatology and leprosy, Rajiv Gandhi Government General hospital were enrolled for the study.

DATA COLLECTION

Based on the proforma, detailed analysis was done which include the history taking, with particular emphasis on the age of onset of the disease, progression of the disease, involvement beyond the palms and soles, discoloration of the keratotic surface, recurrent skin infections, recurrent blisters, constricting bands over digits, auto amputation of

digits, dryness of skin, seasonal variation and collodion baby. History of itching, scaling of the skin, characteristics of scales, pain, fissuring of the keratotic surface, bleeding from the fissures, malodor from the thickened surface, burning sensation, skin lesions elsewhere in the body, pigmentary disturbances and photosensitivity was noted

History of appendageal involvement like hypo or hyperhidrosis, hair changes like woolly hair, teeth involvement, mucosal involvement and nail changes was also recorded. History of involvement of other structures like eyes and ears and involvement of other systems like cardiovascular system, central nervous system was also enquired.

Previous treatment history and response to the treatment were also recorded.Detailed family history, history of consanguinity and involvement of siblings and first degree relatives were noted. Pedigree chart was drawn for patients with hereditary palmoplantar keratoderma.

History suggestive of acquired etiology of Palmoplantar keratoderma like infections, drug intake, reactive dermatoses, systemic illness, hypothyroidism and internal malignancies was recorded.

History suggestive of complications of PPK like pain, fissures, bleeding from the fissures, contractures and secondary infections were noted.

To asses the quality of life, patients were enquired about

1. Itchy, sore, painful or stinging of the skin
2. Embarrassment or self conscious because of skin condition
3. Interference with shopping or looking after home or garden
4. Influence over the clothes
5. Affection over social or leisure activities,
6. Participation in sports
7. Problem at work or studying
8. Any problem in mixing with friends and relatives
9. Sexual difficulties
10. Treatment related problems

Based on the answers, the score were given as “very much” (score 3), “a lot” (score 2), “a little” (score 1) or “not at all” (score 0)“not relevant” (score 0). The maximum score is 30 and the minimum is 0. Based on the score, quality of life was graded as0–1 no effect on patient’s life,2–5 small effect on patient’s life, 6–10 moderate effect on

patient’s life, 11–20 very large effect on patient’s life,21–30 extremely large effect on patient’s life.

Vital signs were recorded. Systemic examination was done methodically. A careful and detailed dermatological examination of palms, soles, other skin areas, hair, nail and mucosa were done. Documentation with clinical photographs was also done. Referral to other specialties (ENT, ophthal, dental, cardiology and neurology) were done as and when required to find out associated features of syndromes.

Complete haemogram, renal function tests, liver function tests, lipid profile, serological test for syphilis, scraping for mite infestation and superficial fungal infections were done. Skin biopsy of hyperkeratotic lesion and biopsy of other related lesions were also done.

ANALYSIS

All the data were analysed and based on that observations were noted.

OBSERVATION AND RESULTS

Out of 86,429 patients who attended our outpatient department during the study period, total number of patients with palmoplantar keratoderma was129 corresponding to 0.15%.

PREVALENCE OF PPK (TABLE-1)

Total no. of new cases	86,429
Total no. of PPK	129
Prevalence of PPK	0.15%

Based on the age of onset, family history, consanguinity and clinical presentation, the patients were classified into hereditary and acquired PPK, consisting of 70 and 59 patients respectively, represents 0.08% and 0.06% of total PPK.

RELATIVE PROPORTION OF EACH CATEGORY (TABLE-2)

Hereditary PPK	70	54.26%	0.08%
Acquired PPK	59	45.73%	0.06%

ANALYSIS OF HEREDITARY PPK

Age distribution

Hereditary PPK constituted 22.85% in the first decade. More commonly seen in the second decade and decreased in prevalence as age advanced.

(TABLE-3)

AGE IN DECADES	HEREDITARY
1-10	16 (22.85%)
11-20	26 (37.14%)
21-30	20 (28. 57 %)
31-40	2 (2.85 %)
41-50	4 (5.71%)
51-60	2 (2.85%)

SEX DISTRIBUTION

In the gender wise analysis, males constituted a higher proportion in hereditary PPK (61.42%)

(TABLE-4)

DISEASE	MALE	FEMALE	TOTAL
Hereditary	43 (61.42%)	27 (38.57%)	70 (54.29%)

OCCUPATION

In hereditary PPK, students were the majority people (42.85%), followed by unemployed people (18.57%).

(TABLE-5)

OCCUPATION	HEREDITARY
Student	30 (42.85%)
Unemployed	13(18.57%)
Laborer	9 (12.85%)
House wife	7 (10%)
Office work	3 (4.2%)
Engineer	2 (2.8%)
Fisherman	2 (2.8%)
Painter	1 (1.4%)
Weaver	1 (1.4%)

According to the clinical pattern, the common pattern was diffuse nontransgrediens type, males constituted more than 2/3 patients with this type. Among females(14.2%) diffuse transgrediens types is most prevalent

(TABLE-6)

Clinical pattern	No. of cases	Male	Female
Diffuse-transgreidiens	15 (21 %)	05 (7.1 %)	10 (14.2 %)
Diffuse–nontransgrediens	48(68.57 %)	29(4 %)	19(30 %)
Focal	6 (7.1 %)	6(7.1 %)	Nil
Punctate	1(1.4 %)	1(1.4 %)	Nil

Out of the 70 patients with hereditary palmoplantar keratoderma, 43 patients were found to have palmoplantar keratoderma syndromes,27 patients were found to have PPK associated with other genodermatosis.

Different types of PPK syndromes and their prevalence is shown in the following table. Unna Thost syndrome was the common one with prevalence of 30%.

(TABLE-7)

S.NO	Clinical Types	No. of Cases	HPPK
1	Unna Thost syndrome	21	30%
2	Mal de Meleda	4	5.71%
3	Naxos disease	2	2.85%
4	Griethers disease	3	4.28%
5	Olmsted syndrome	3	4.28%
6	Papillon Lefevere syndrome	2	2.85%
7	Clouston syndrome	1	1.42%
8	Focal plantar keratoderma	6	8.57%
9	Marginal keratoderma	1	1.42%

Hereditary PPK found associated with other genodermatosis totally constituted about 38.57% of hereditary PPK

(TABLE-8)

S. No	Genodermatosis	No. of cases	Percentage
1	Ichthyosis Vulgaris	6	8.57%
2	Bullous Icthyosiform Erythroderma	3	4.28%
3	Lamellar ichthyosis	3	4.28%
4	Sjogren Larsson syndrome	1	1.42%
5	Familial pitryasis rubra pilaris	6	8.57%
6	Kindler syndrome	4	5.71%
7	Epidermolysis bullosa	1	1.42%
8	Symmetrical progressive erythrokeratoderma	1	1.42%
9	Pachy dermo periosteitis	1	1.42%
10	Dowling Degos disease	1	1.42%

Family history and consanguinity in relation to hereditary PPK 52.85% of patients had positive family history of similar illness. 3^0 consanguinity was noted in 47.14% patients

(TABLE-9)

Consanguinity	Positive family history	Negative family history
Non consanguinity	4	22
2^0 consanguinity	6	5
3^0 consanguinity	27	6

Since Unna Thost syndrome constituted about 30% of hereditary PPK, we have analysed it separately. It was commonly seen in males.

(TABLE-10)

Total no cases	Male	Female
21	15	6

Unna Thost syndrome & Consanguinity

Out of the 21 patients with Unna Thost syndrome,9 had positive family history and 12 had negative family history.9 patients born out of nonconsanquinous marriage, 1 from 2^0 Consanguinity and 11 from 3^0 Consanguinity.

(TABLE-11)

Consanguinity	Positive family history	Negative family history
Non consanguinity	1	8
2^0 consanguinity	Nil	1
3^0 consanguinity	8	3

Dermatological Life Quality Index (DLQI)

Hereditary PPK had a significant impact on life. 22.85 % of people had an extremely large effect on life.47.14% had a very large effect on life.

(TABLE-12)

S.NO	CATEGORY	No. cases in HPPK
1	No effect on life	Nil
2	Small effect on life	9(12.85 %)
3	Moderate effect on life	12(17.14 %)
4	Very large effect on life	33(47.14 %)
5	Extremely large effect	16(22.85 %)

ANALYSIS OF ACQUIRED PPK

Acquired PPK is commonly found in the 5th decade of life(33.89%) followed by the 3rd decade. No case was seen in the first decade.

(TABLE-13)

AGE IN DECADES	ACQUIRED
1-10	Nil
11-20	6 (10.16 %)
21-30	9 (15.25%)
31-40	8 (13.55%)
41-50	20 (33.89%)
51-60	8 (13.55%)
61-70	3 (5.08%)
71-80	4 (6.77%)
81-90	1 (1.69%)

SEX DISTRIBUTION

The prevalence of acquired PPK was more in females.

(TABLE-14)

DISEASE	MALE	FEMALE	TOTAL
Acquired	27(45.76%)	32 (54.23%)	59(45.73%)

OCCUPATION

Here housewives and manual laborers topped the list (33.89%) followed by students with 10.1%

(TABLE-15)

OCCUPATION	ACQUIRED PPK
House wife	20 (33.89%)
Laborer	20 (33.89%)
Unemployed	6 (10.1%)
Student	5 (8.7%)
Fisherman	4 (6.7%)
Mechanic	2 (3.3%)
Office work	2 (3.3%)

CLINICAL PATTERN

Focal type of keratoderma was the most common presentation of acquired PPK with slight male preponderance.

(TABLE-16)

Clinical pattern	Total no cases	Male	Female
Diffuse	27 (45.7%)	10(16.97%)	17(28.87 %)
Focal	32(54.27%)	17(28.87%)	15(25.47 %)
Punctate	Nil	Nil	Nil

ETIOLOGY

Out of the 59 patients, psoriasis constituted about 64.40 %, followed by hyperkeratotic hand and foot eczema.

(TABLE-17)

S.No	Disease	Total no cases	Percentage
1	Psoriasis	38	64.40%
2	Eczema	8	13.55%
3	Pityriasis rubra pilaris	4	6.77%
4	Erythroderma dermatitis	2	3.38%
5	Crusted scabies	2	3.38%
6	Reiters syndrome	1	1.16%
7	Mycosis fungoides	1	1.16%
8	Atopic dermatitis	1	1.16%
9	Lichen planus	1	1.16%
10	Pitted keratolysis	1	1.16%

PSORIASIS

Among the patients with psoriasis, females outnumbered the males. It was commonly seen in the fifth decade.

(TABLE-18)

No. of cases	Male	Female
38	16(27.11%)	22(37.28%)

Dermatological Life Quality Index (DLQI)

Out of those suffering from the acquired PPK ,64.4% of people had very large effect on life.

(TABLE-19)

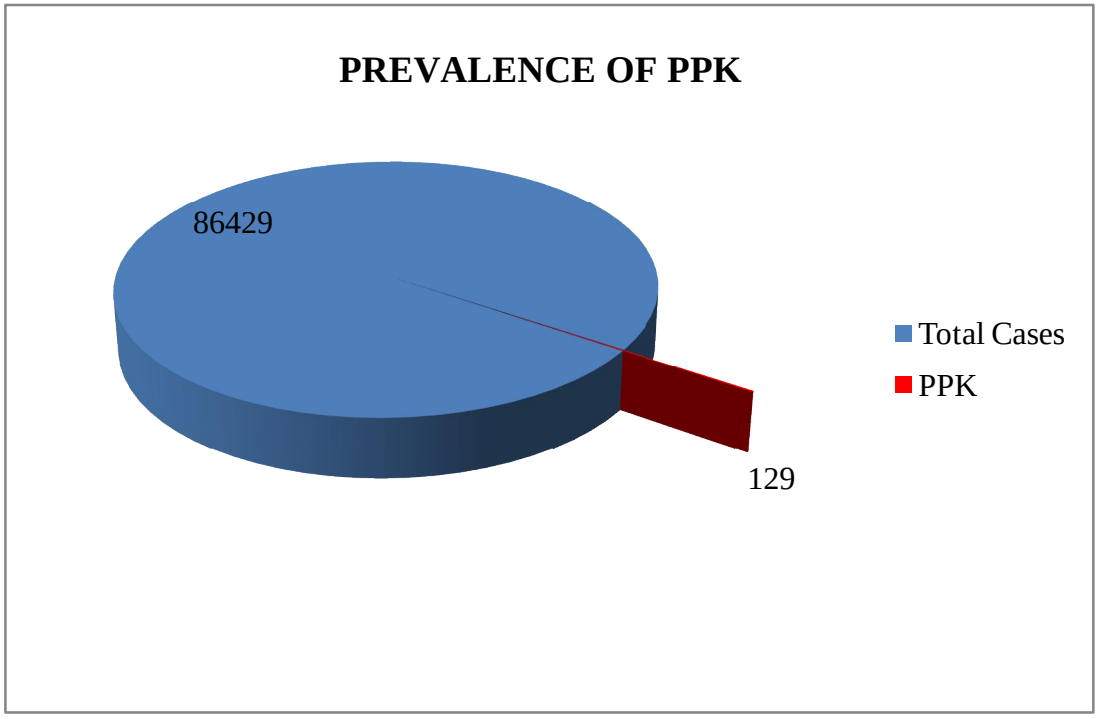
S.No	Category	No. cases in acquired PPK
1	No effect on life	Nil
2	Small effect on life	2 (3.38 %)
3	Moderate effect on life	17 (28.8 %)
4	Very large effect on life	38 (64.4 %)
5	Extremely large effect	2 (3.38 %)

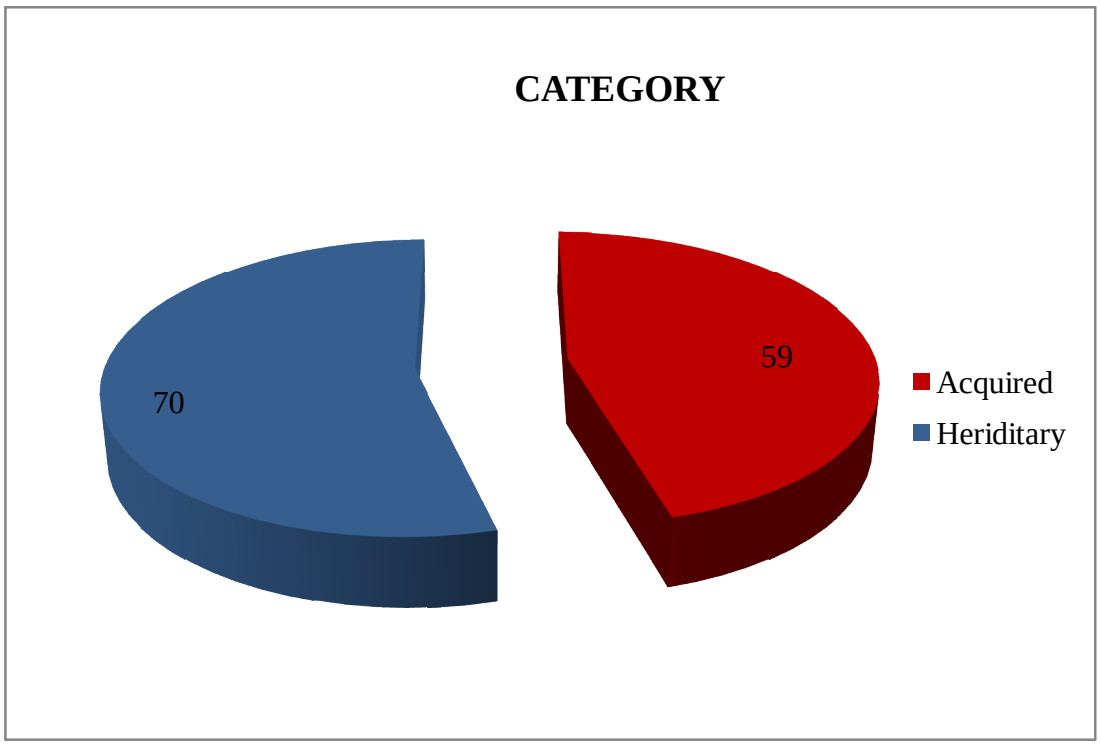
NAIL CHANGES

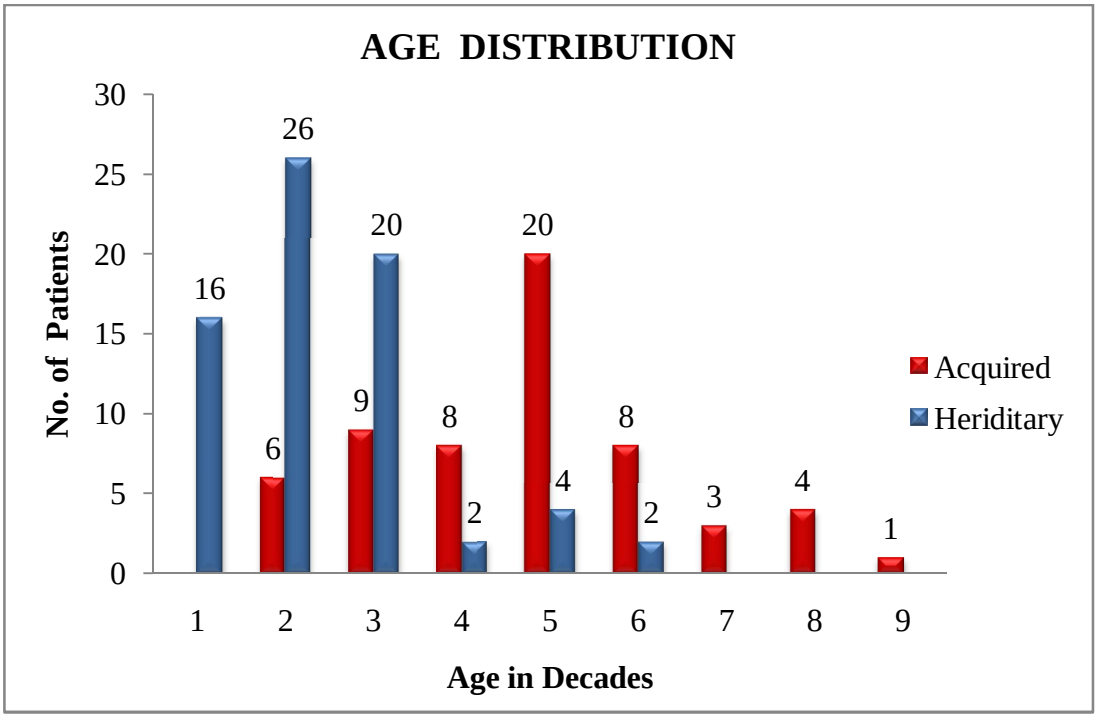
Out of the 129 patients 81 patients had nail changes as shown in bar chart. Most common change was pitting.

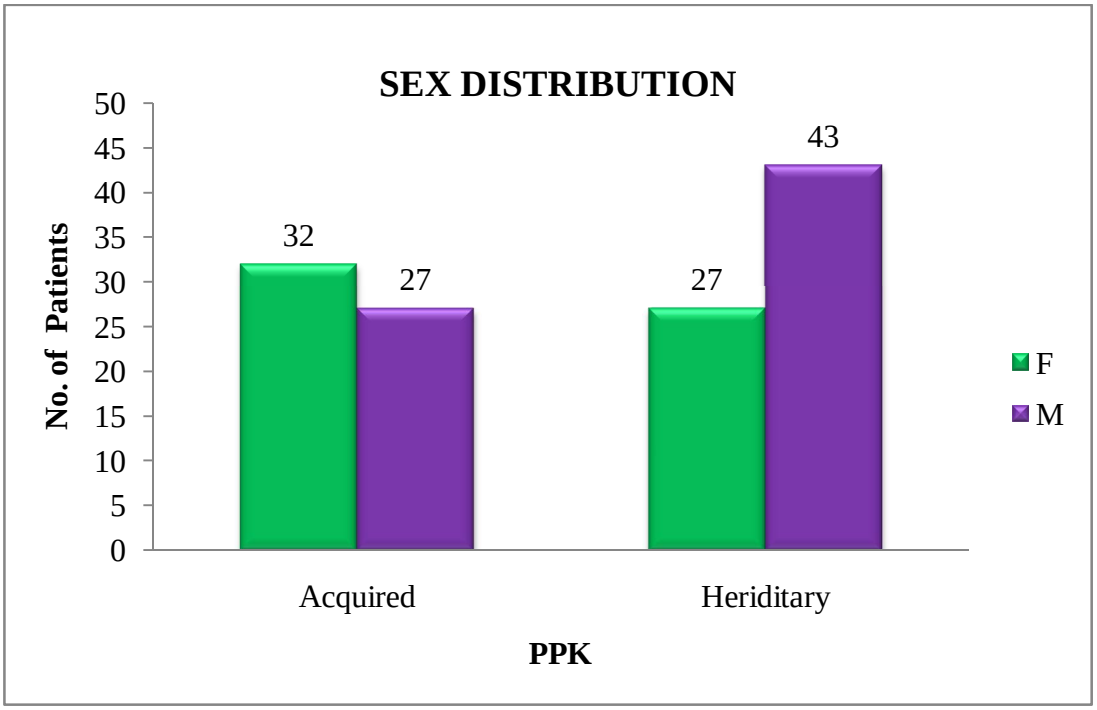
COMPLICATIONS

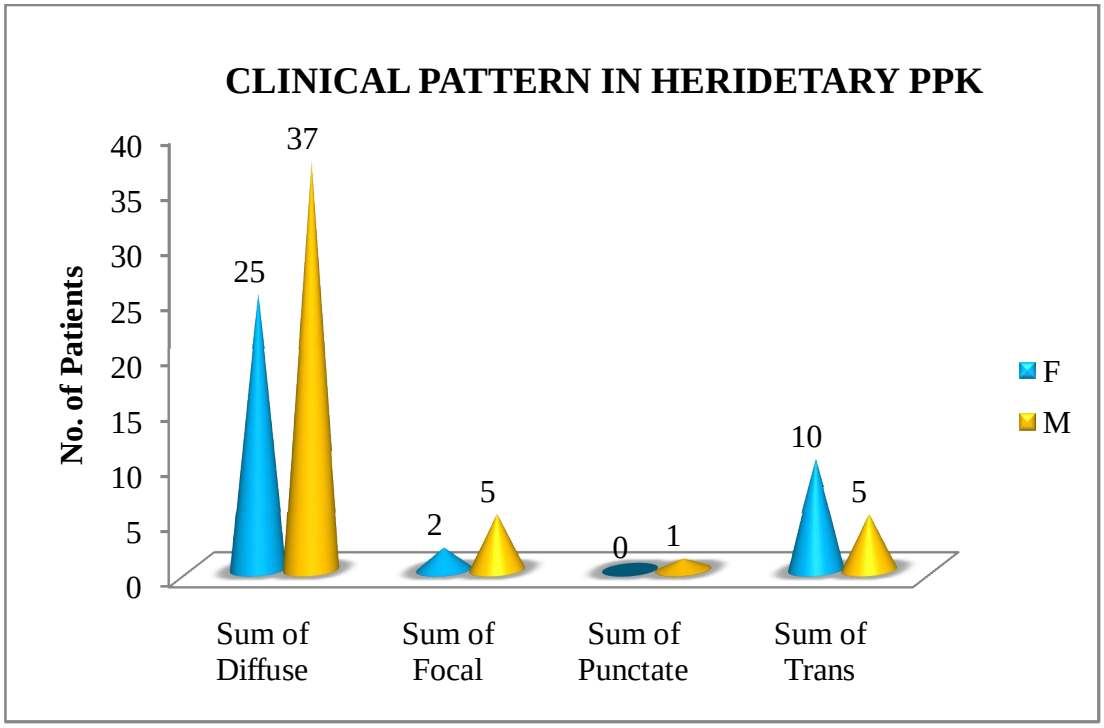
Complications are mostly seen in hereditary PPK than acquired PPK. The most common one observed was fissuring, followed by contractures and bleeding from fissures and pain. Secondary skin infection was also observed, among which pitted keratolysis was common. This may due to hyperhidrosis and occupation related. Pyodermas and malodour was also observed. Autoamputation of digit due to pseudoainhum was observed in one patient.

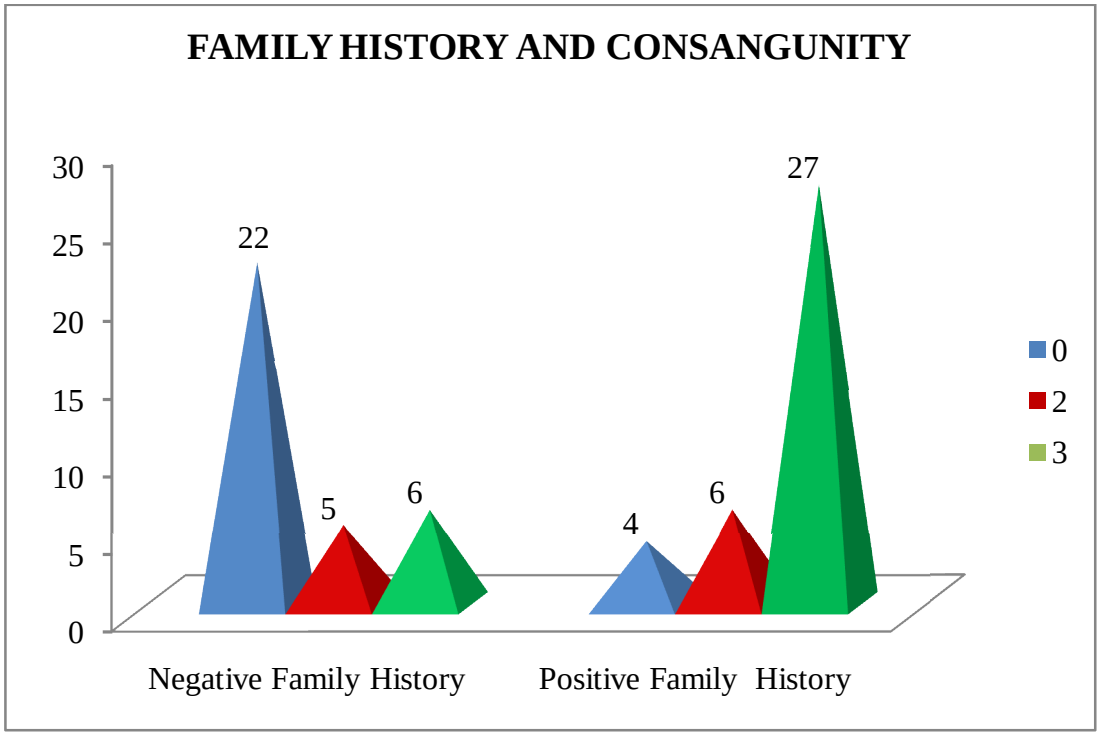

PREVALENCE OF PPK
86429
129
Total Cases
PPK

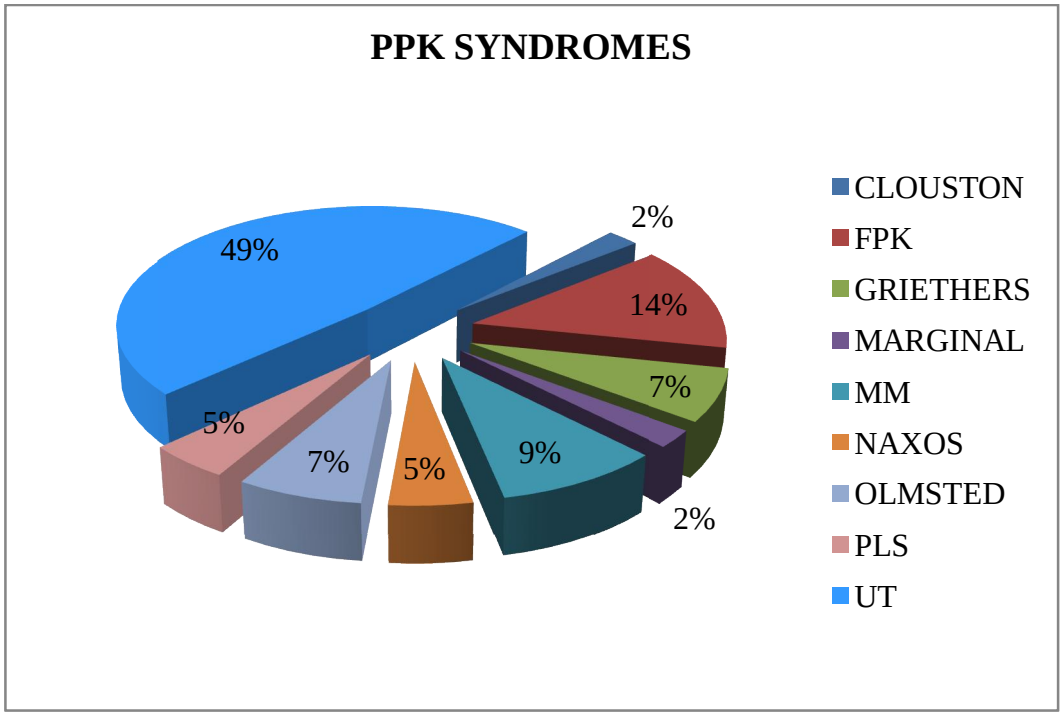

CATEGORY
70
59
Acquired
Heriditary

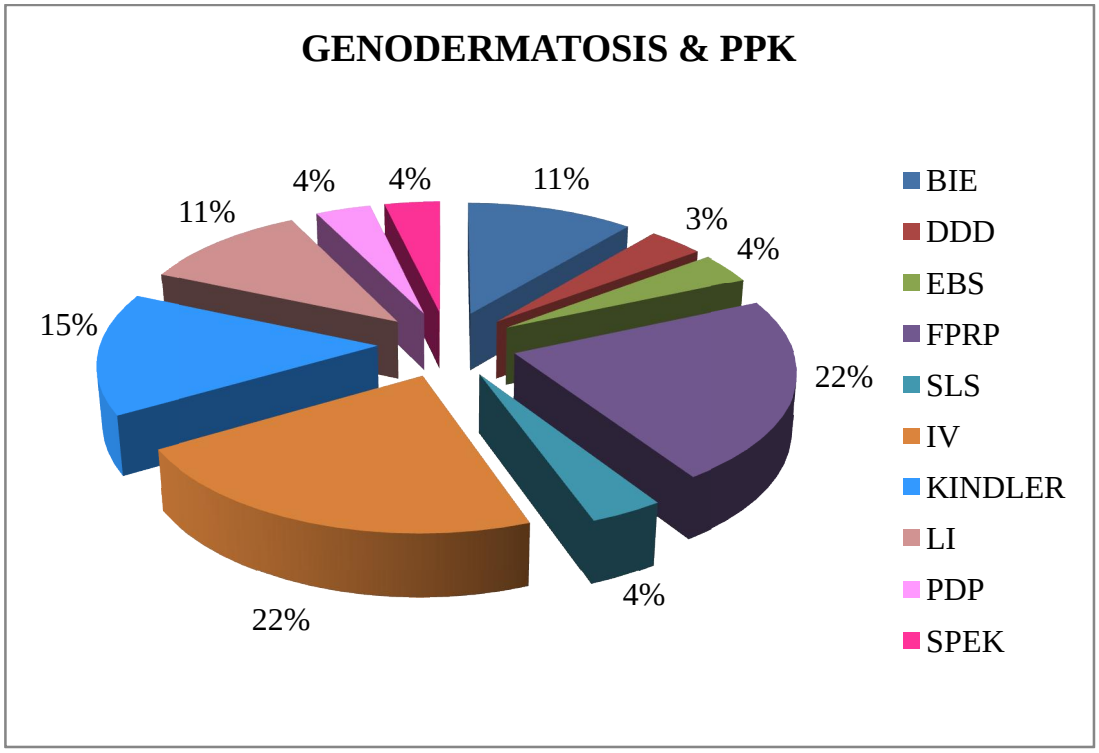

AGE DISTRIBUTION
No. of Patients
30
25
20
15
10
5
0
16
26
6
20
9
8
2
20
4
8
2
3
4
1
1
2
3
4
5
6
7
8
9
Age in Decades
Acquired
Heriditary

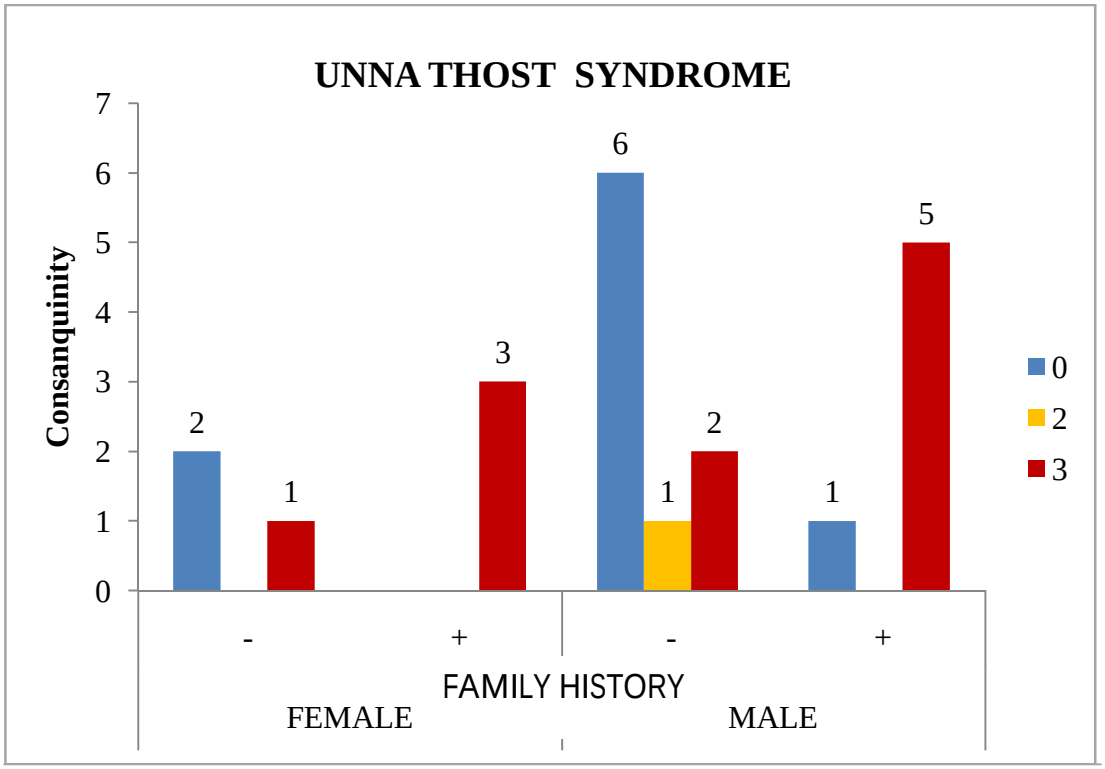

SEX DISTRIBUTION
No. of Patients
50
45
40
35
30
25
20
15
10
5
0
32
27
27
43
Acquired
Heriditary
PPK
F
M

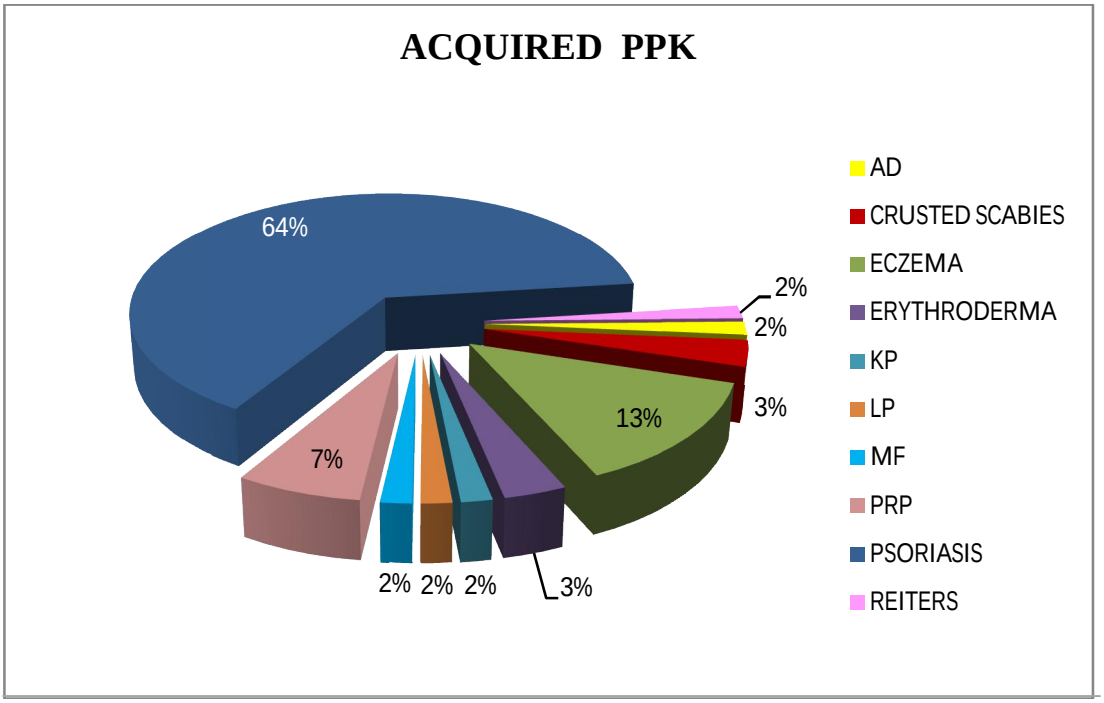

CLINICAL PATTERN IN HERIDETARY PPK
No. of Patients
40
35
30
25
20
15
10
5
0
25
37
2
5
0
1
10
5
Sum of Diffuse
Sum of Focal
Sum of Punctate
Sum of Trans
F
M

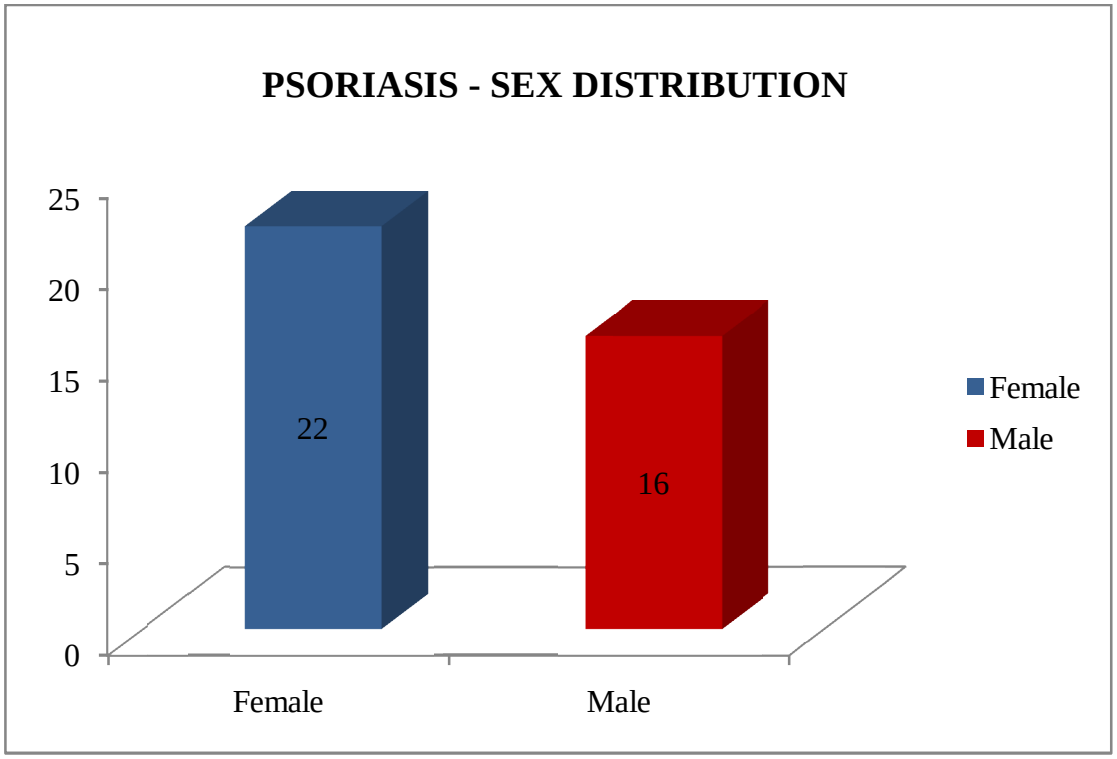

FAMILY HISTORY AND CONSANGUNITY
30
25
20
15
10
5
0
22
5
6
4
6
27
Negative Family History
Positive Family History
0
2
3

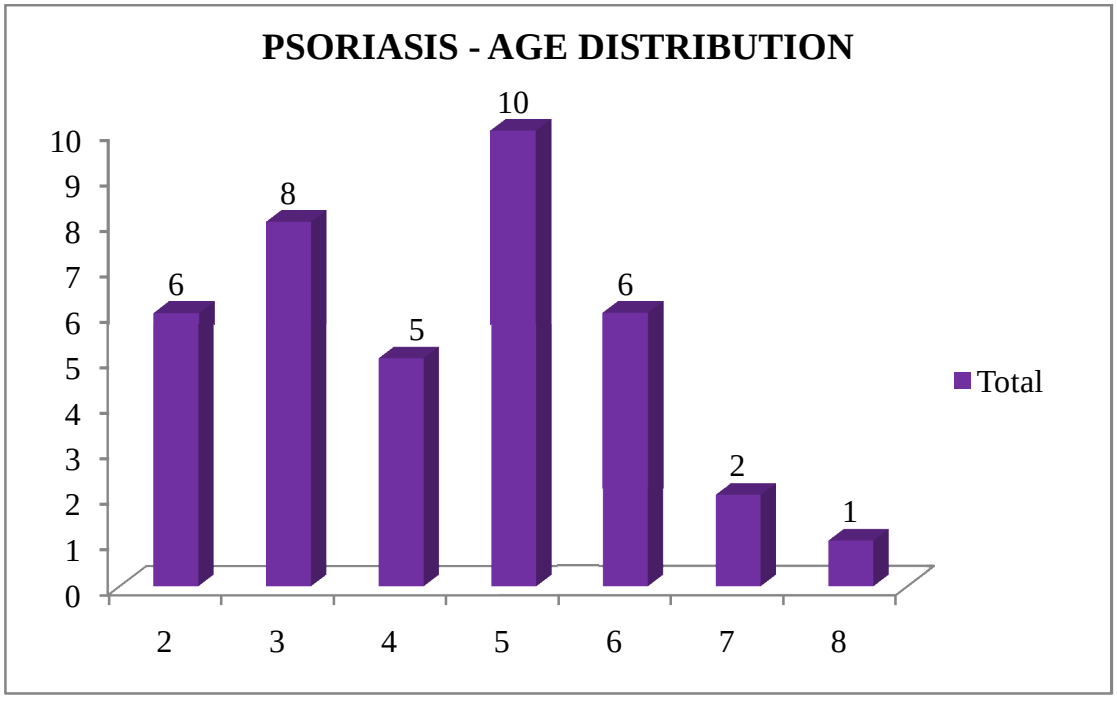

PPK SYNDROMES
49%
2%
14%
7%
5%
7%
5%
9%
2%
CLOUSTON
FPK
GRIETHERS
MARGINAL
MM
NAXOS
OLMSTED
PLS
UT

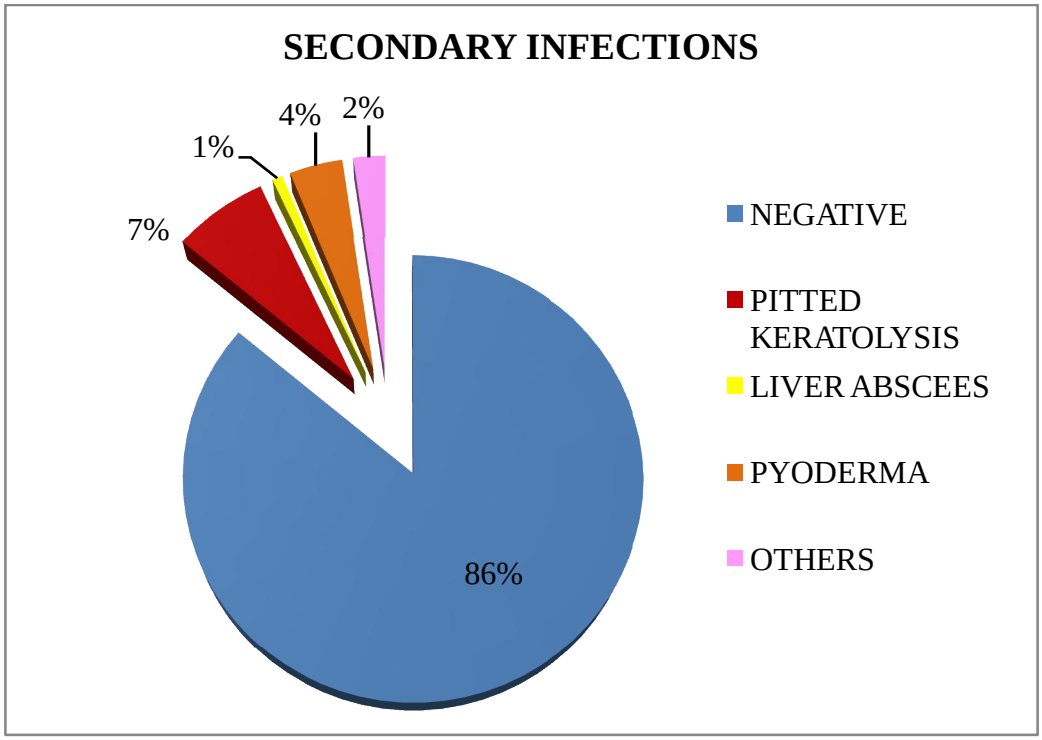

GENODERMATOSIS & PPK
11%
3%
4%
22%
4%
22%
15%
11%
4%
4%
BIE
DDD
EBS
FPRP
SLS
IV
KINDLER
LI
PDP
SPEK

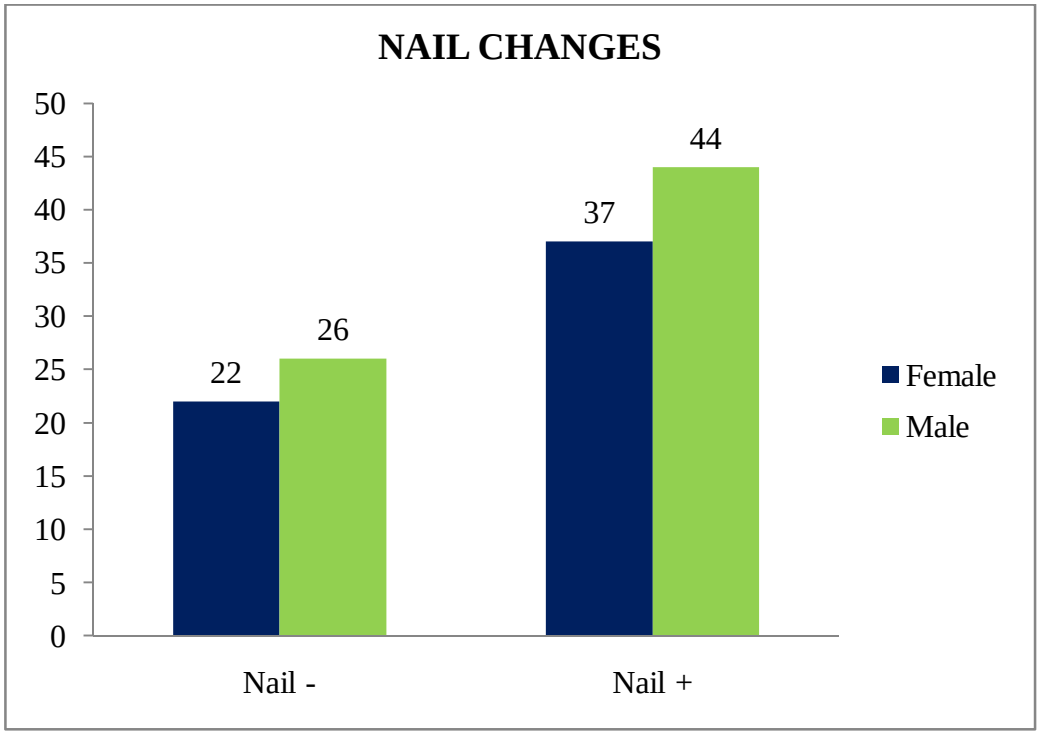

UNNA THOST SYNDROME
Consanquinity
7
6
5
4
3
2
1
0
2
1
3
6
1
2
1
5
0
2
3
-
+
-
+
FAMILY HISTORY
FEMALE
MALE


ACQUIRED PPK
64%
2%
2%
3%
13%
7%
2% 2% 2%
3%
AD
CRUSTED SCABIES
ECZEMA
ERYTHRODERMA
KP
LP
MF
PRP
PSORIASIS
REITERS

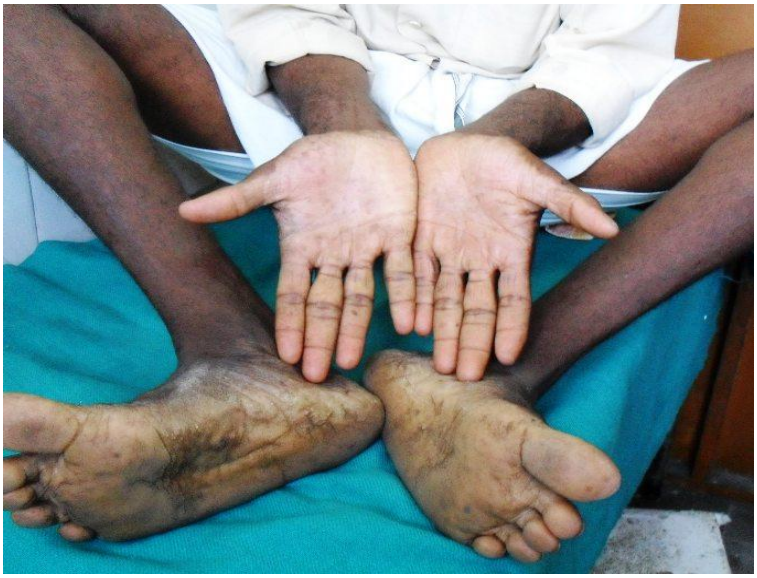

PSORIASIS - SEX DISTRIBUTION
25
20
15
10
5
0
22
16
Female
Male
Female
Male

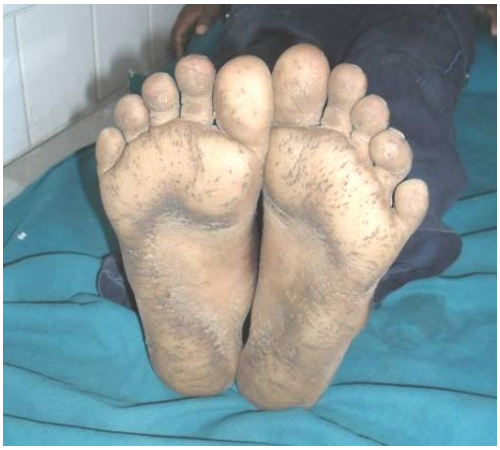

PSORIASIS - AGE DISTRIBUTION
10
9
8
7
6
5
4
3
2
1
0
6
8
5
10
6
2
1
2
3
4
5
6
7
8
Total

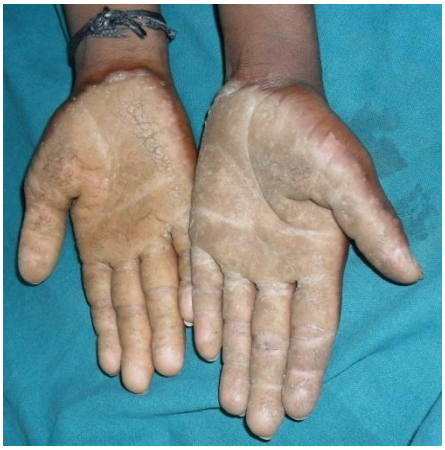

SECONDARY INFECTIONS
4%
2%
1%
7%
86%
NEGATIVE
PITTED KERATOLYSIS
LIVER ABSCEES
PYODERMA
OTHERS


NAIL CHANGES
50
45
40
35
30
25
20
15
10
5
0
22
26
37
44
Nail -
Nail +
Female
Male

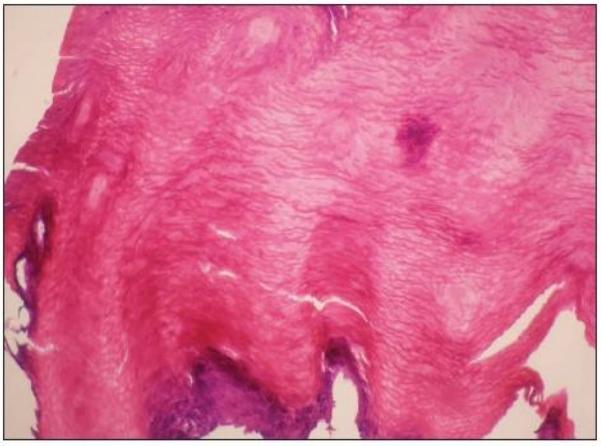
UNNA THOST SYNDROME



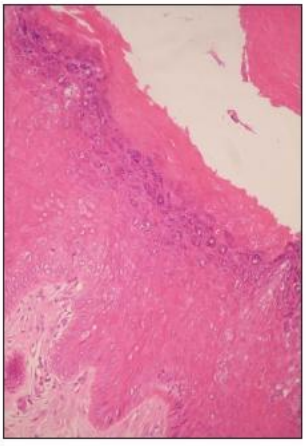




Diffuse PPK

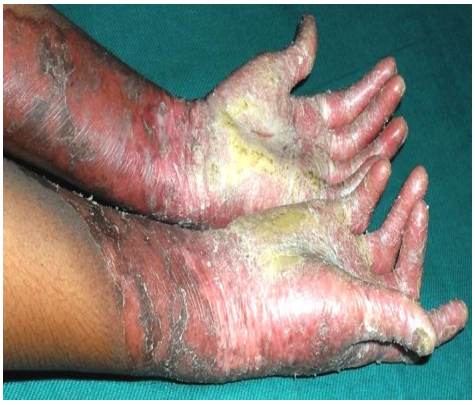


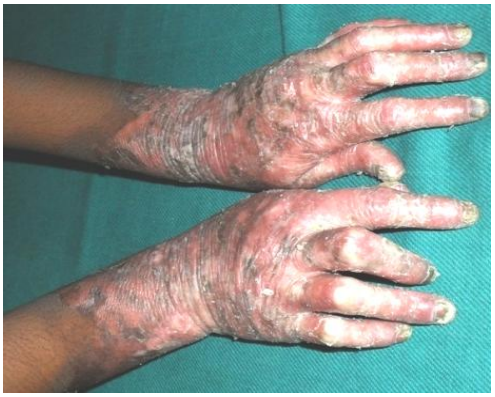
Massive Hyperkeratosis



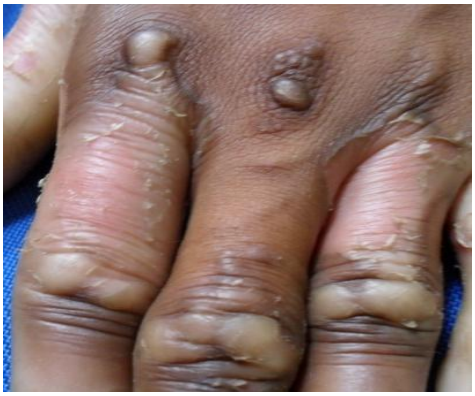
Nonepidermolytic Hyperkeratosis

MAL DE MELEDA

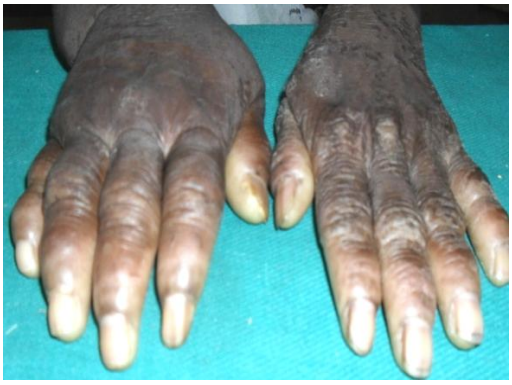




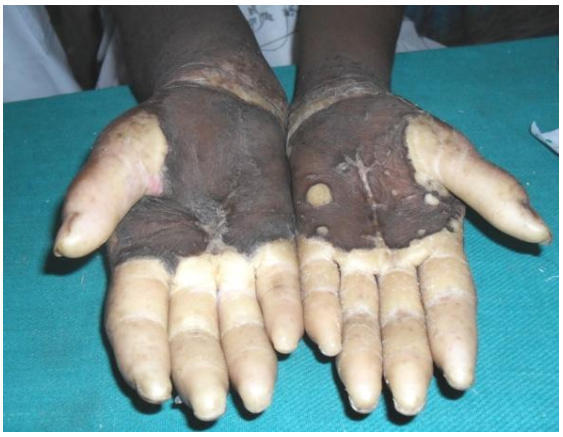
Diffuse Transgrediens



Knuckle Pads

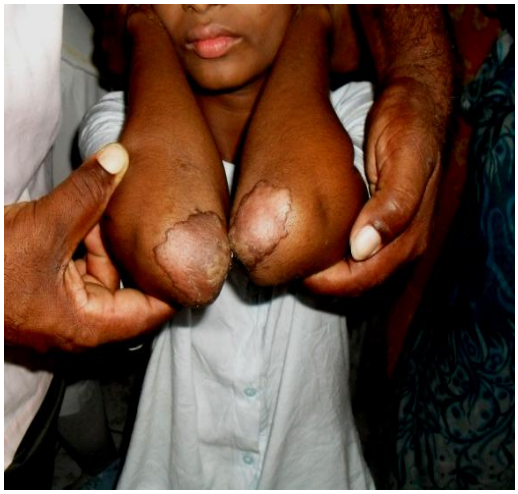


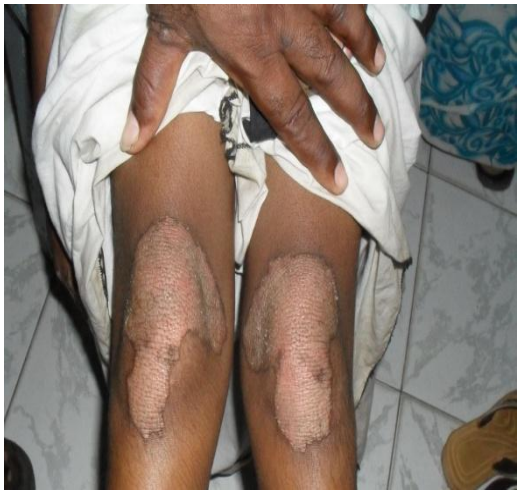
Tapering of Digits



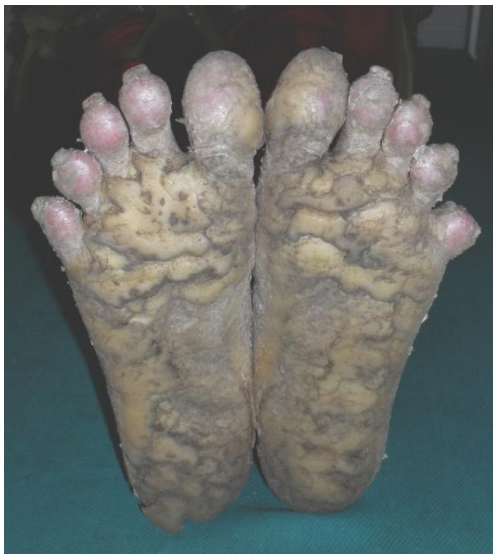
PPK with Skin Grafting

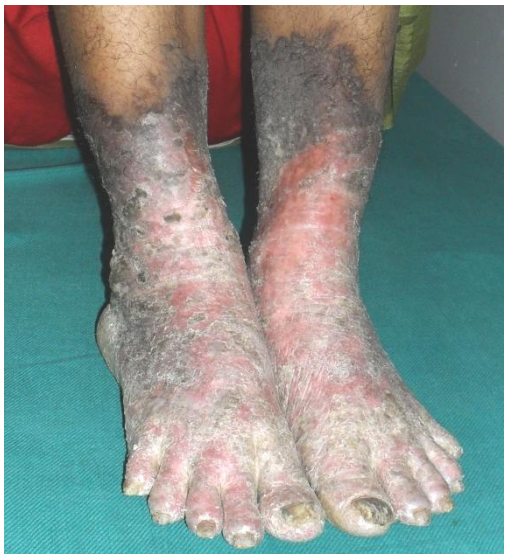
MAL DE MELEDA



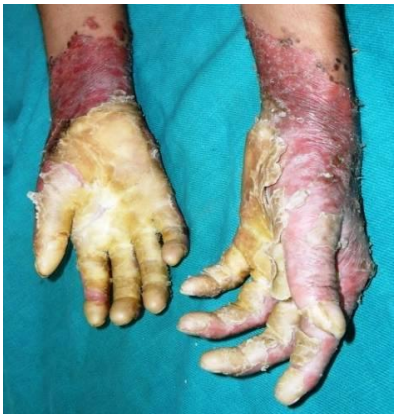


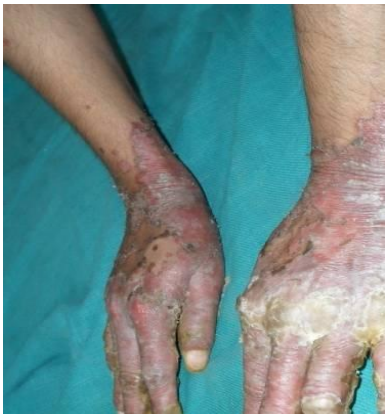
Keradoderma over Elbows & Knees

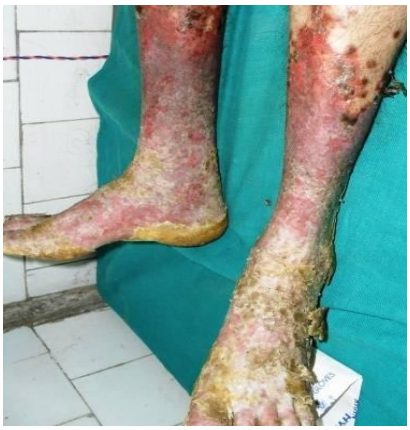


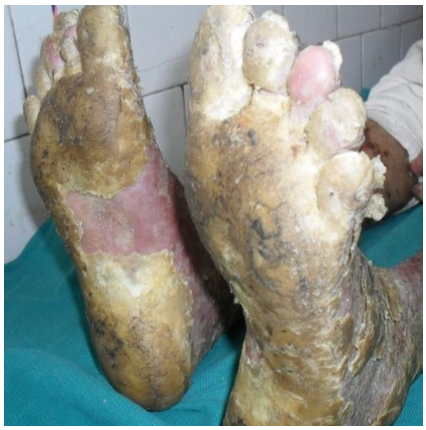


GREITHERS DISEASE IN A FAMILY

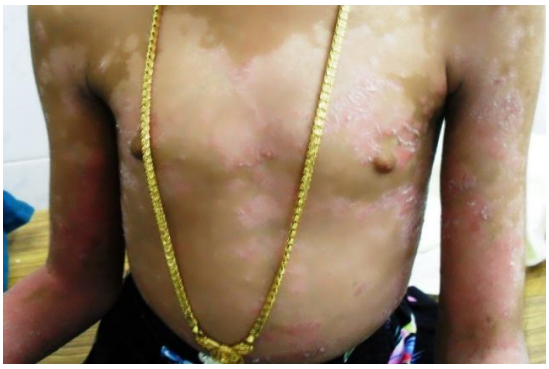


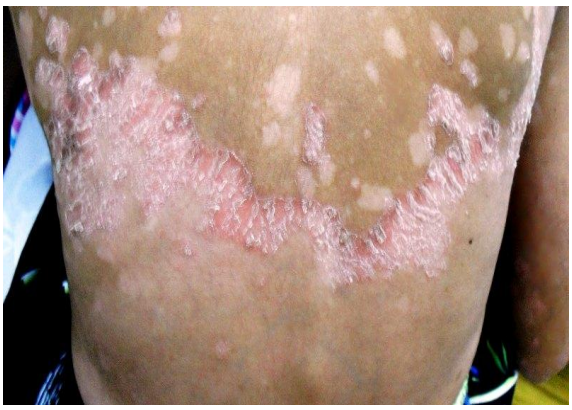


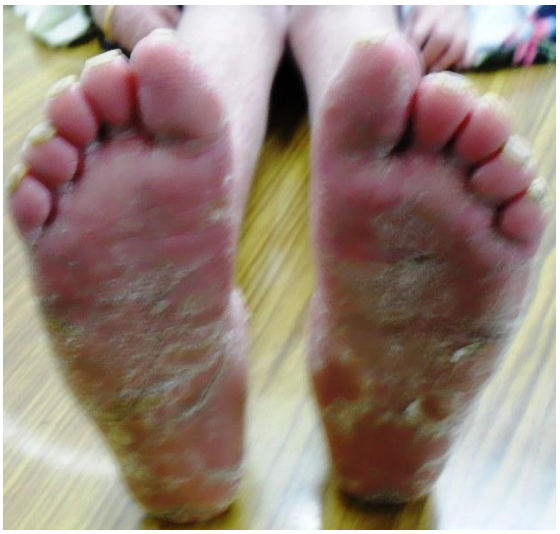




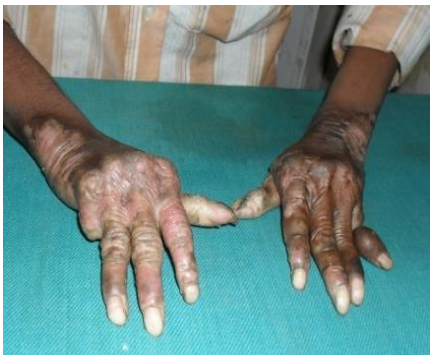
GREITHERS DISEASE INVOLVEMENT UPTO TRUNK



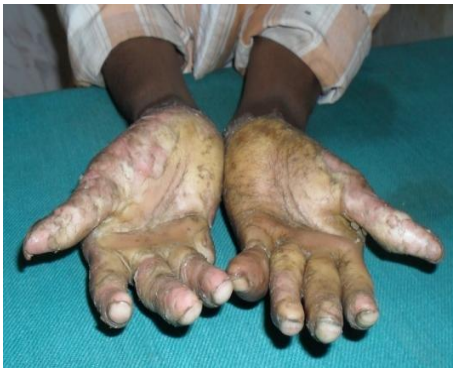


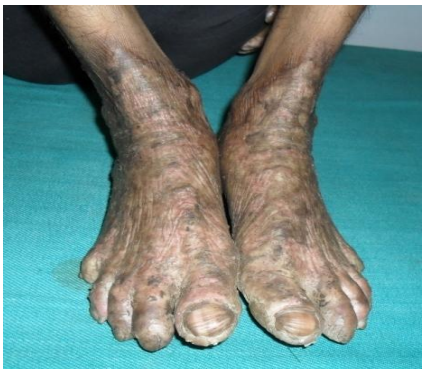


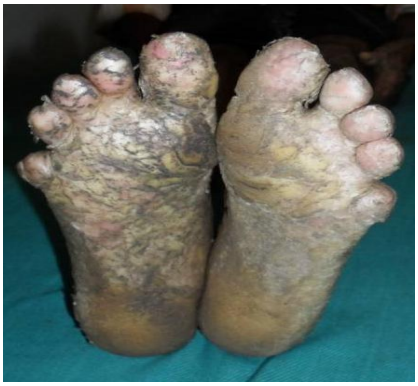
OLMSTED SYNDROME

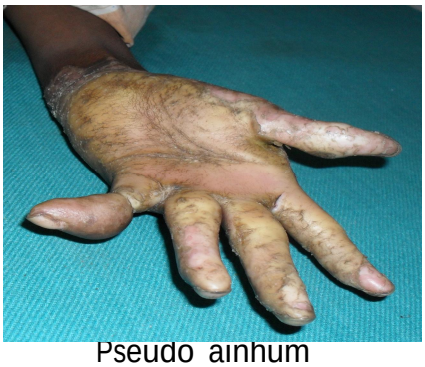


Auto Amputation of Digit



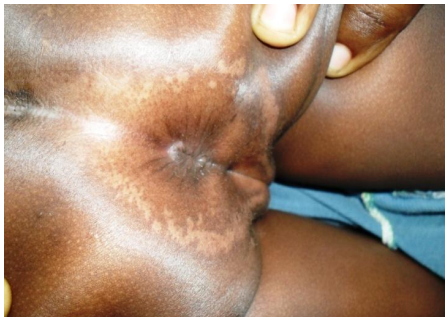




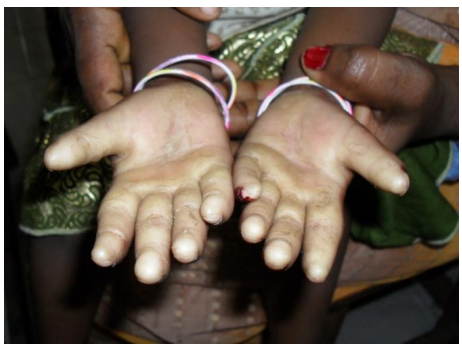


Pseudo ainhum

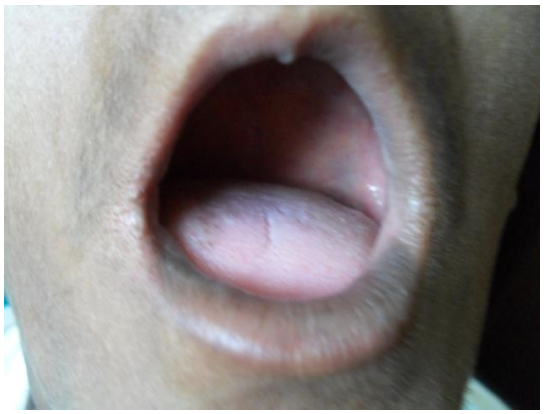
Periorificial thickening



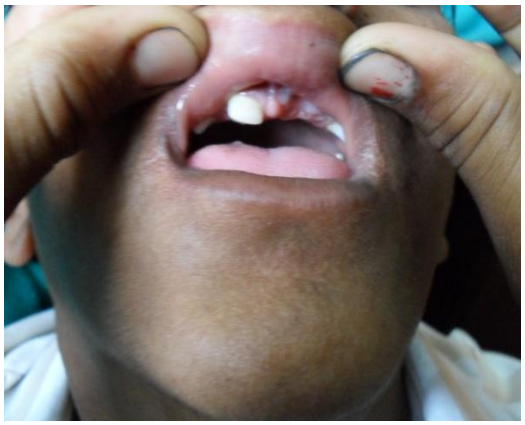
Perianal thickening



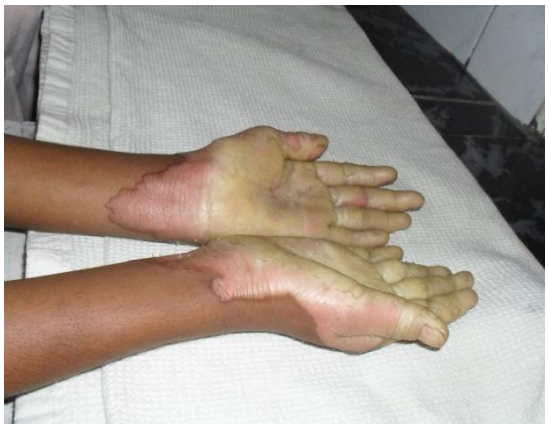
PAPILLON – LEFEVRE S YNDROME



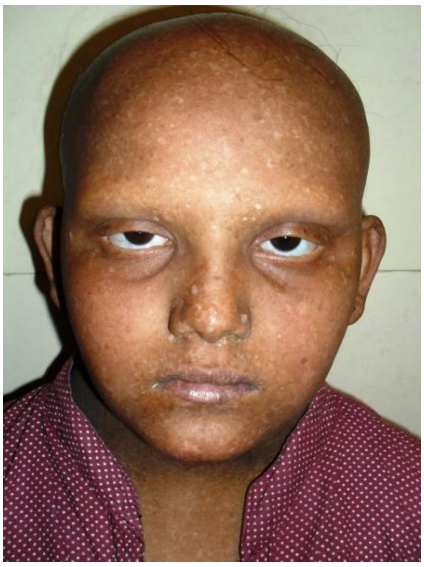
Loss of Teeth

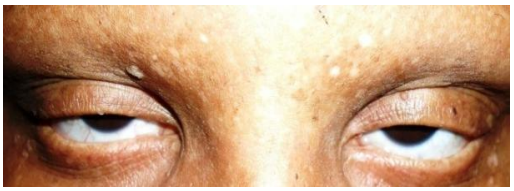


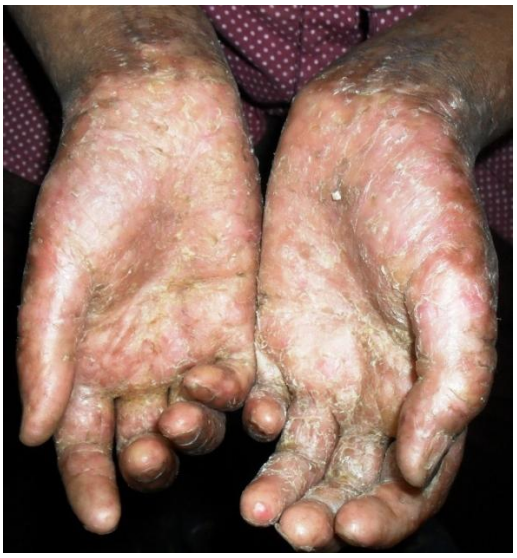
Gingivitis

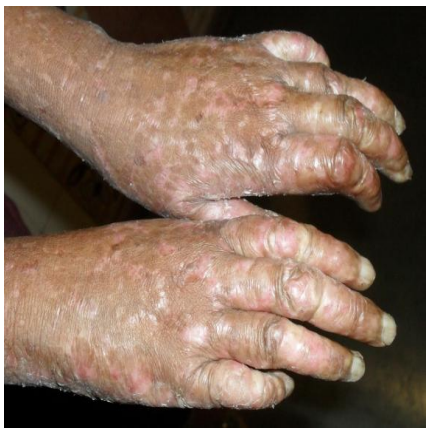


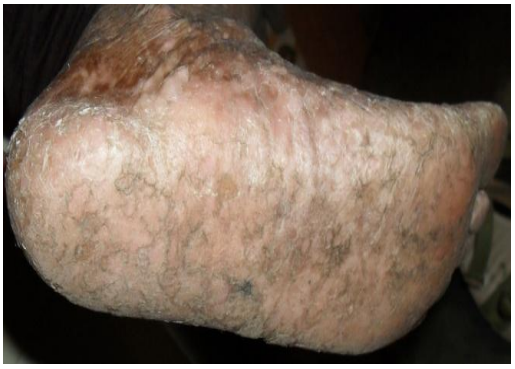
CLOUSTON SYNDROME

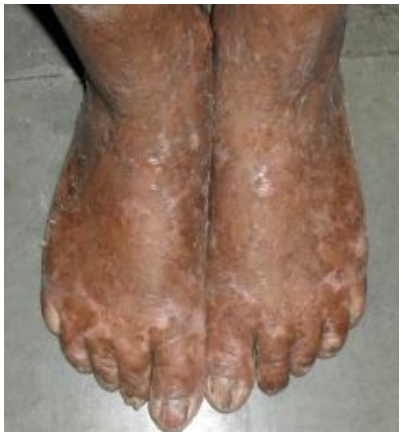




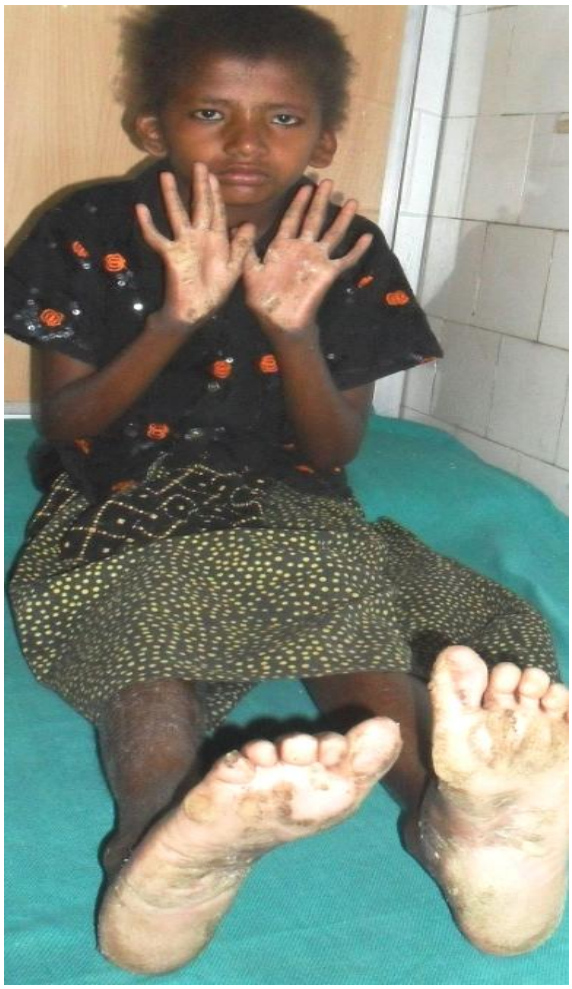


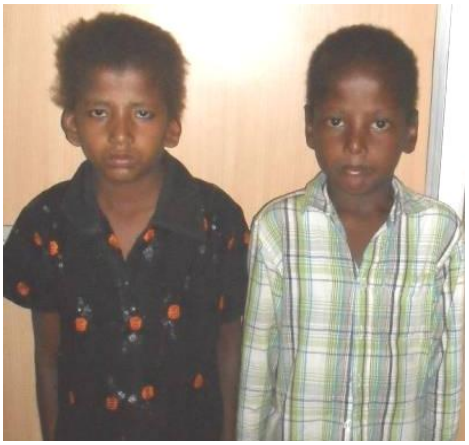


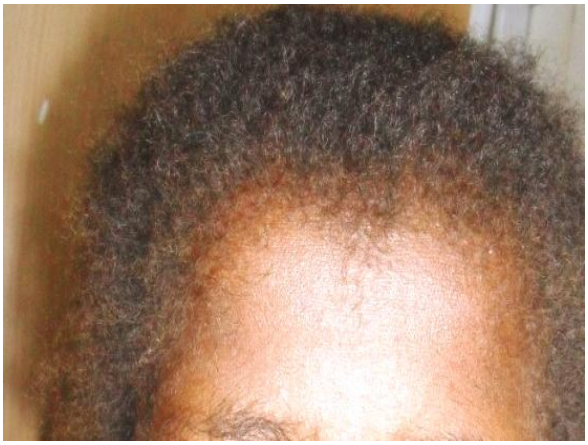




NAXOS DISEASES

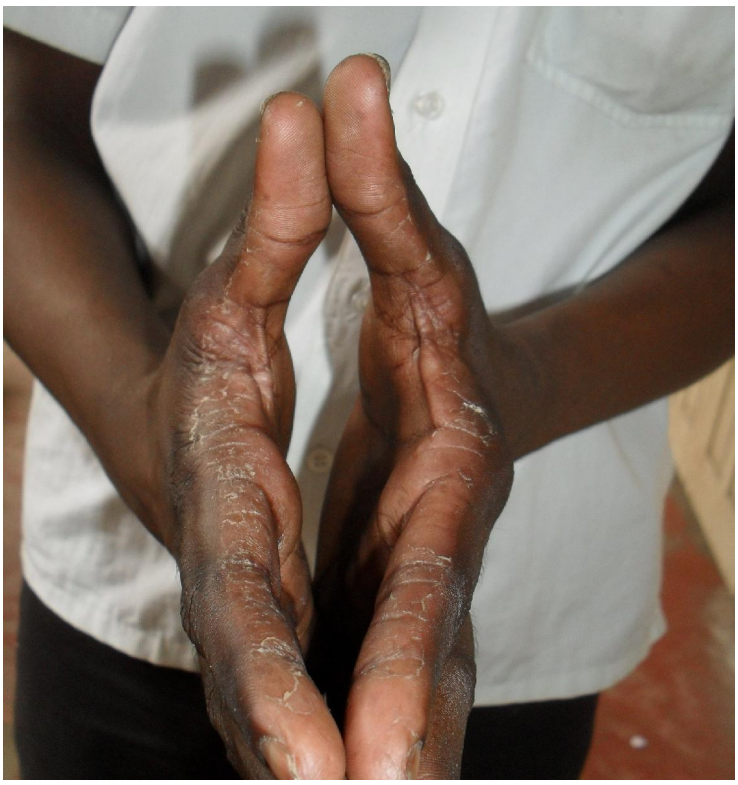




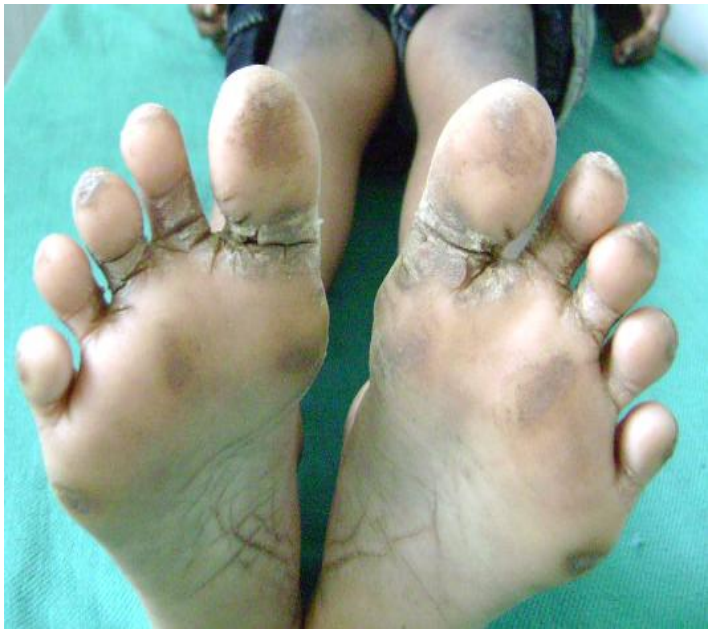


Woolly hair

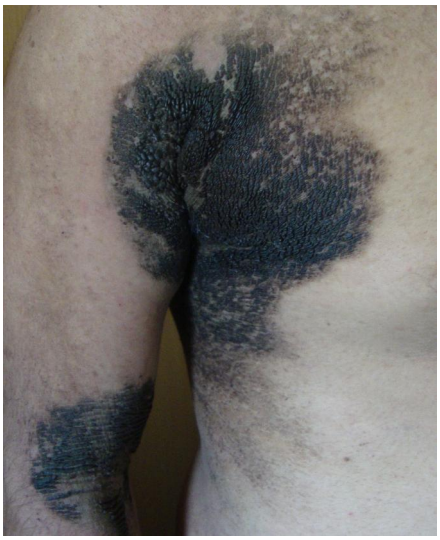
MARGINAL KERATODERMA

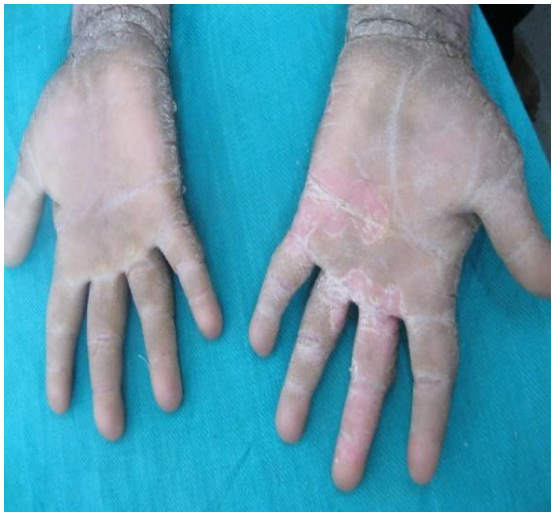


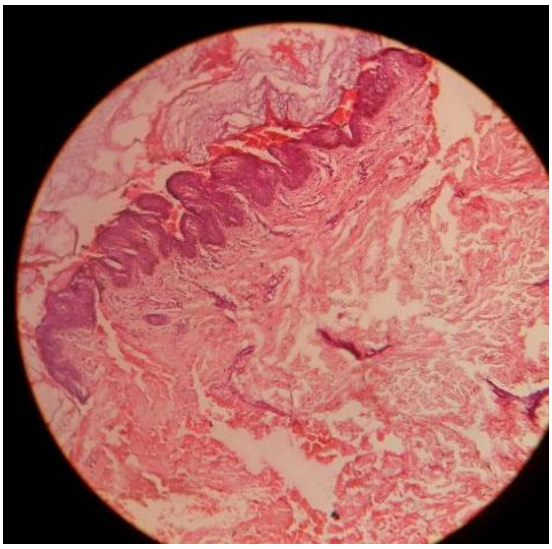
FOCAL PLANTAR KERATODERMA



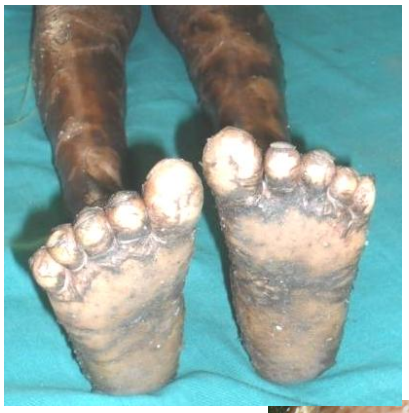
BULLOUS ICHTSIFORM ERYTHRODERMA

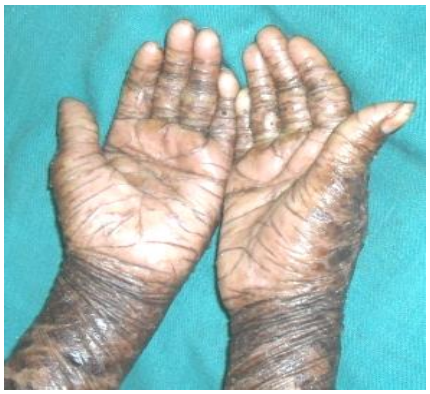


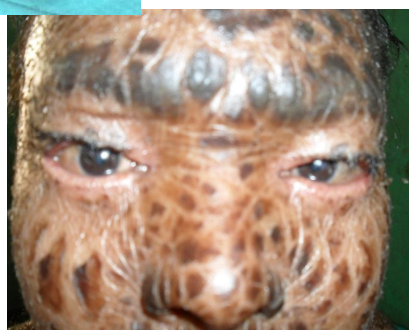




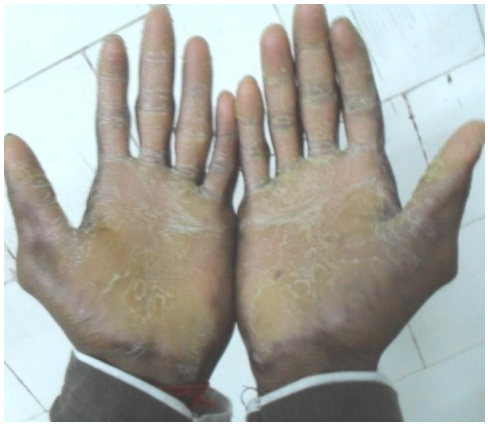
LAMELLAR ICHTHYOSIS WITH PPK

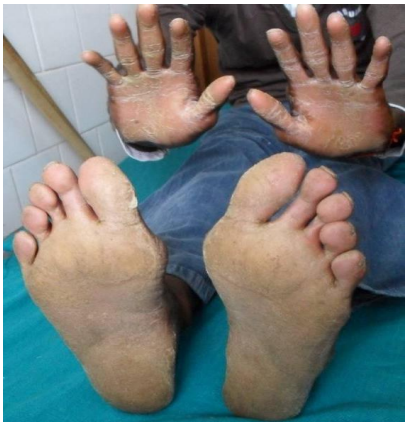


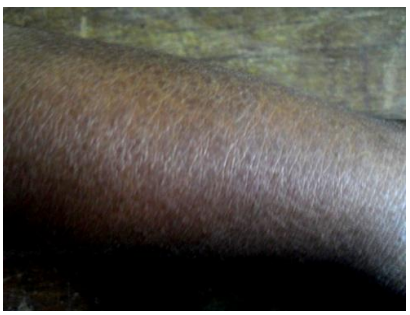




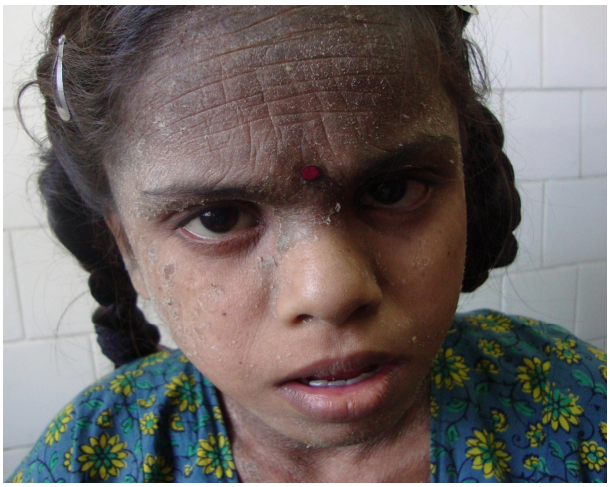
ICHTHYOSIS VULGARIS WITH PPK

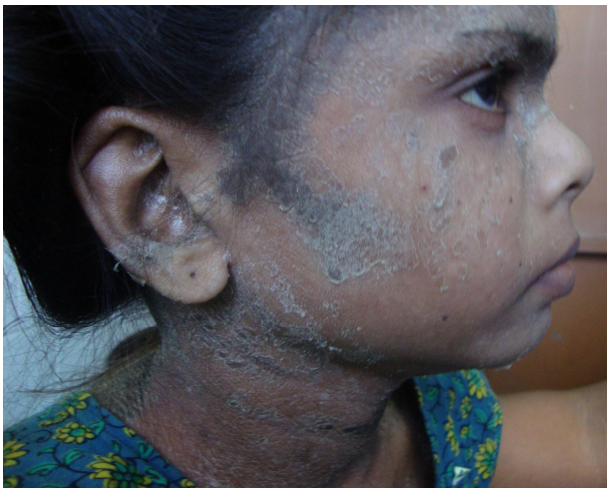


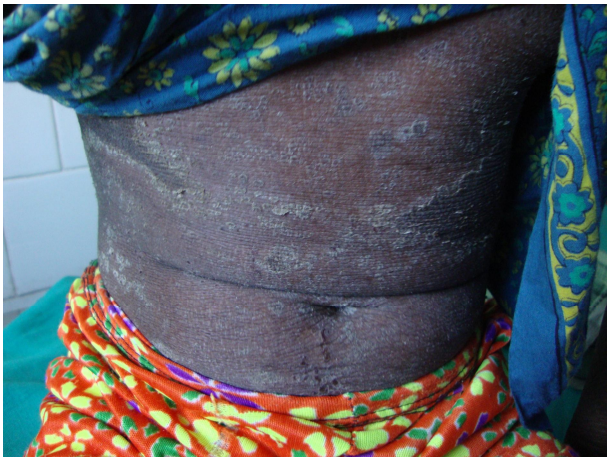




SJOGREN LARSSON SYNDROME

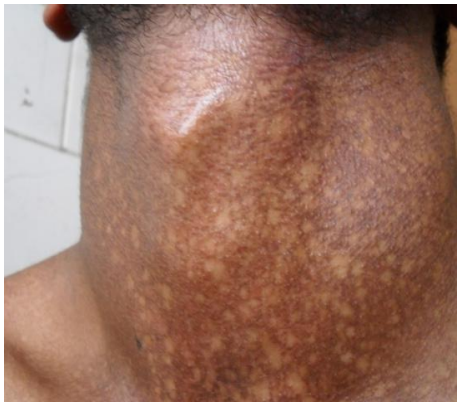




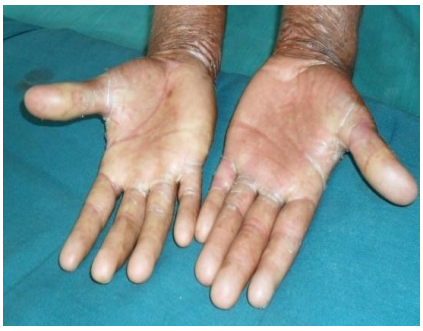


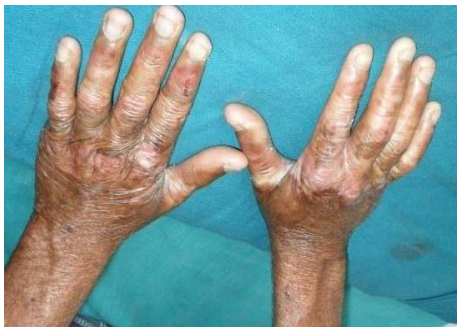
DIFFUSE SKIN PEELING WITH MILD ERYTHRODERMA

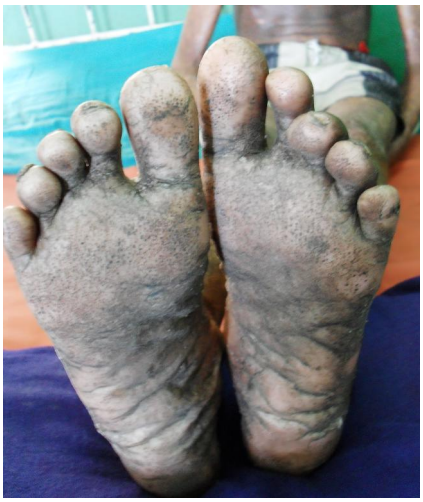
KINDLER SYNDROME

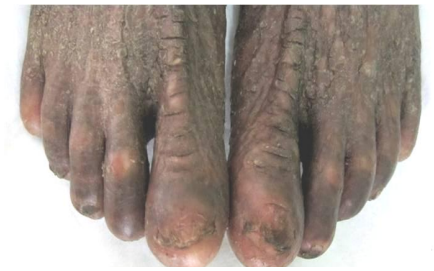


Mottled Pigmentation

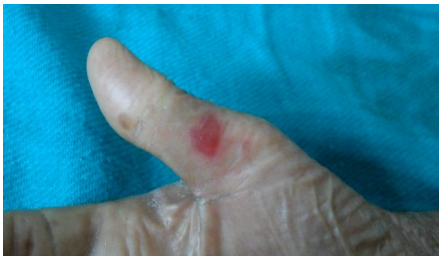




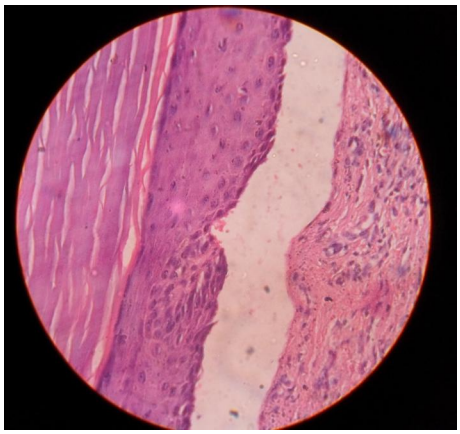




Anonychia of Great Toe Nail

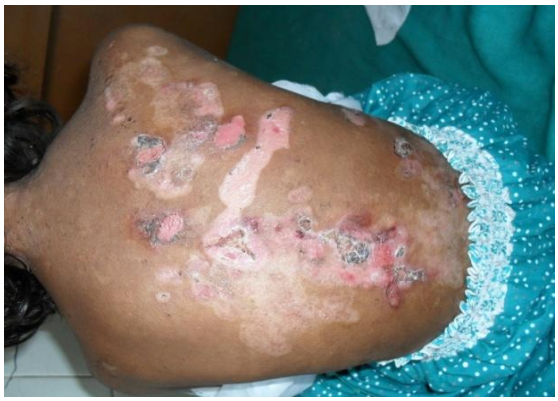


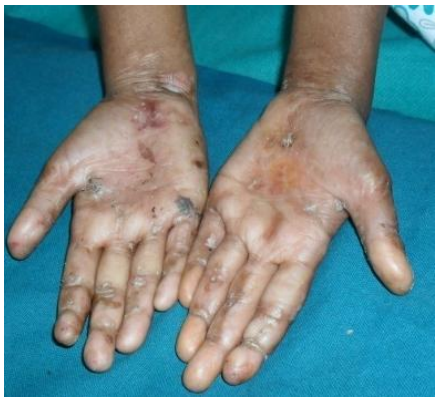
Haemraghic Blister

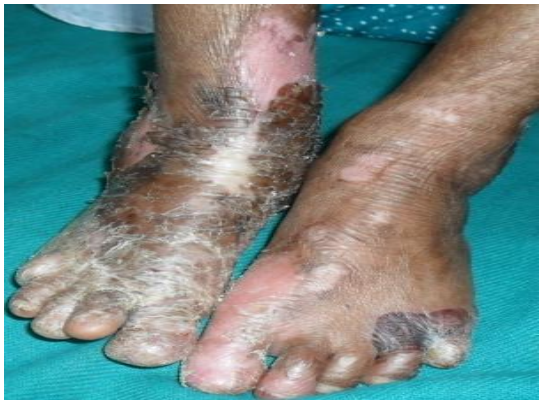


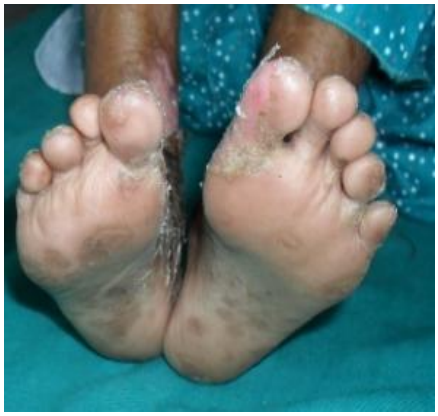
Subepidermal Cleavage

EPIDERMOLYSIS BULLOSA WITH PPK

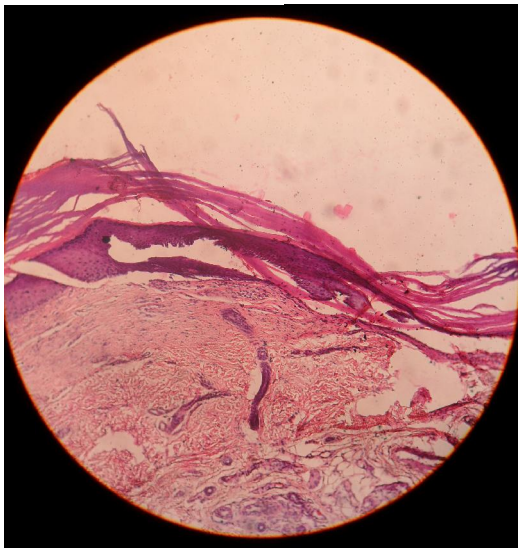






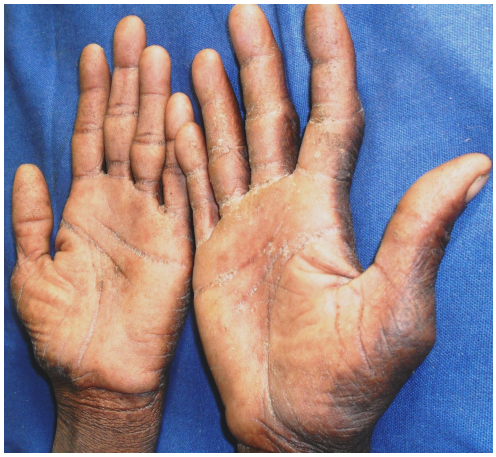


Spontaneous resorption of Rt 5th toe

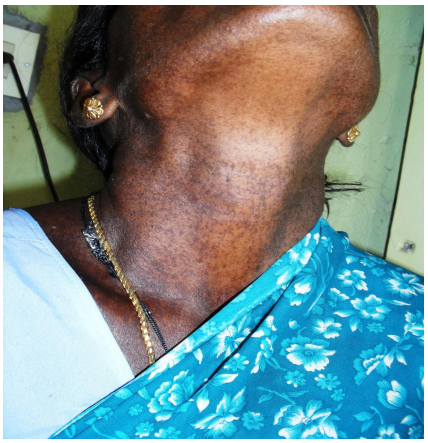


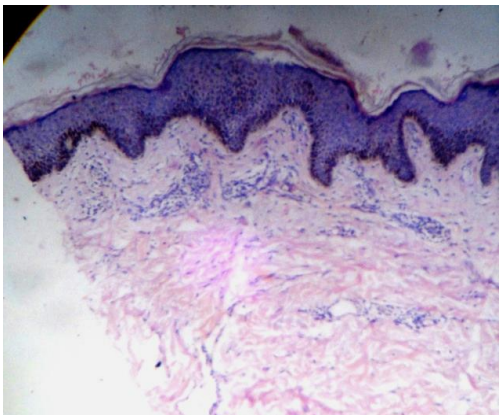
Sub Epidermal Bulla

DOWLING DEGOS DISEASE



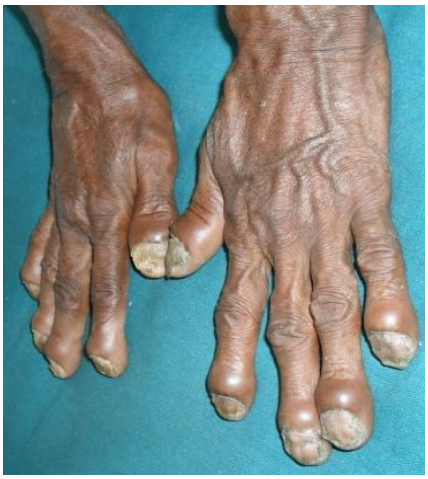
Reticulate Pigmentation

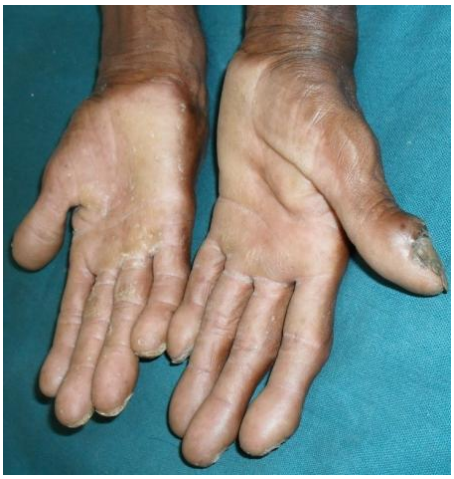


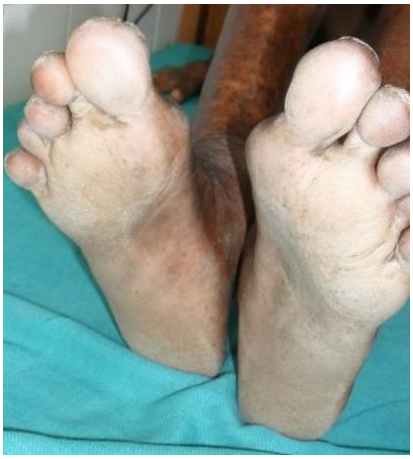


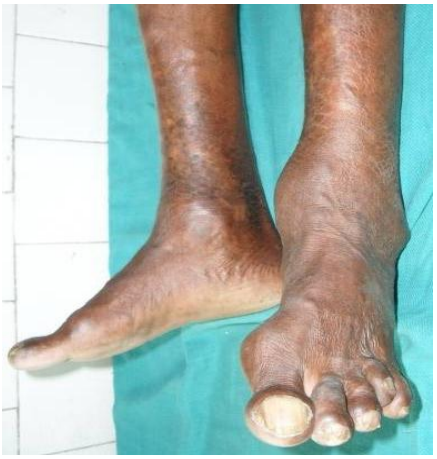
Histopathology

PACHY DERMO PERIOSTEITIS WITH PPK

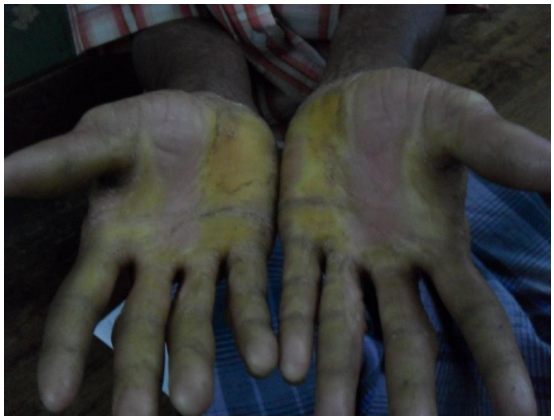
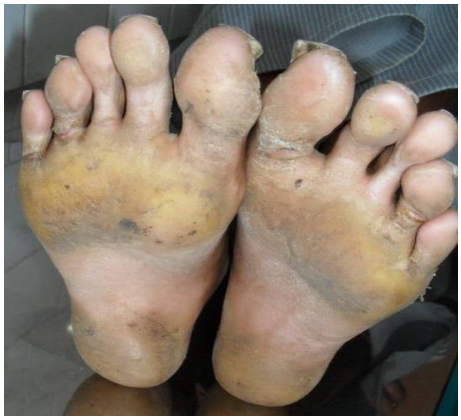




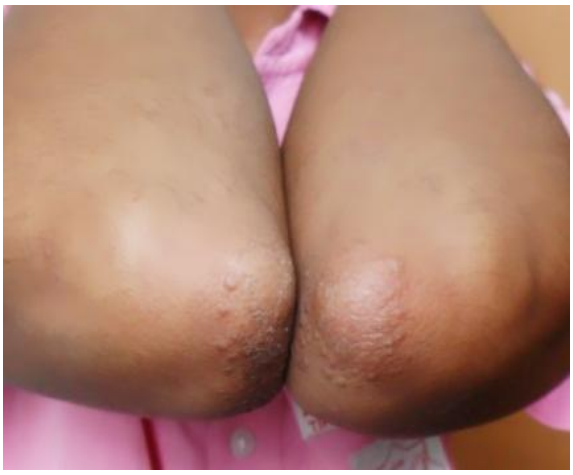
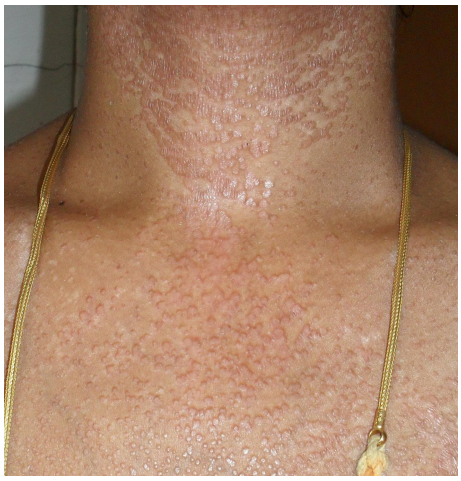


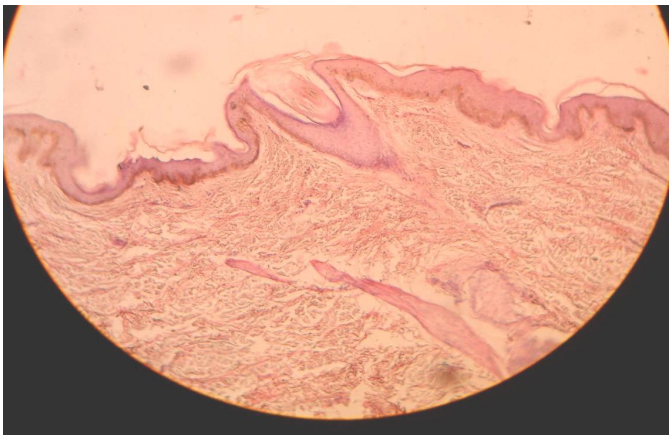


PITYRIASIS RUBRA PILARIS

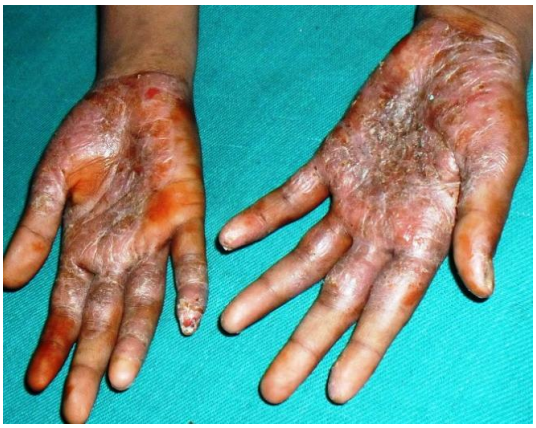
PRP Sandal

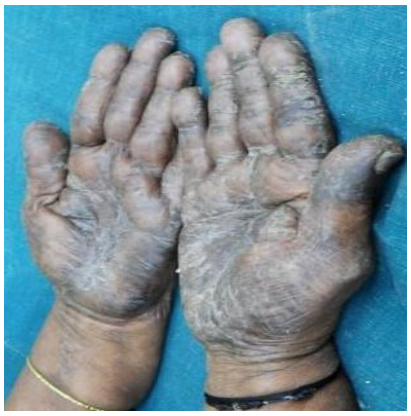





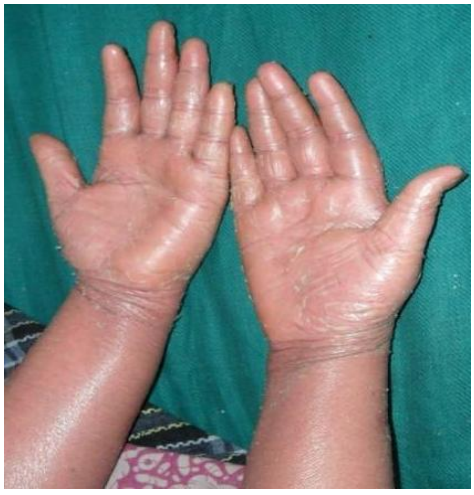
Follicular Plugging with Perifollicular Parakeratosis

PSORIASIS

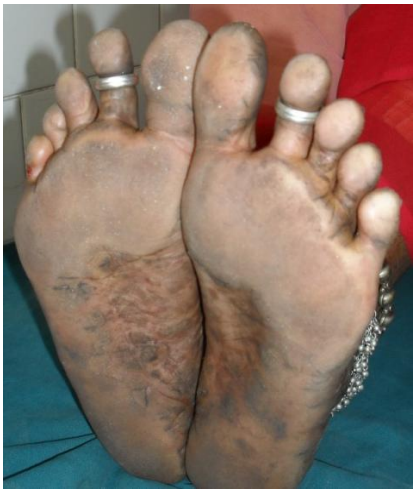




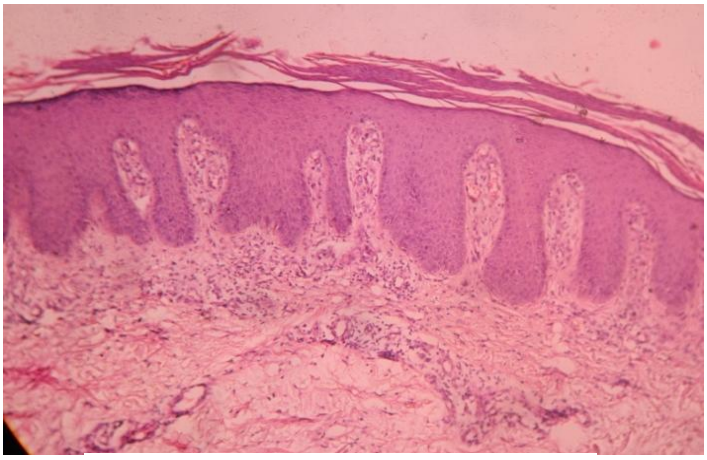
Psoriasis of Palms



Erythrodermic Psoriasis with PPK

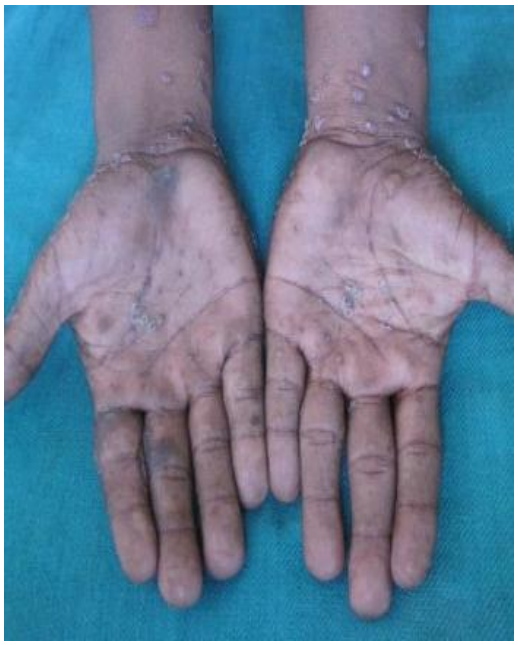


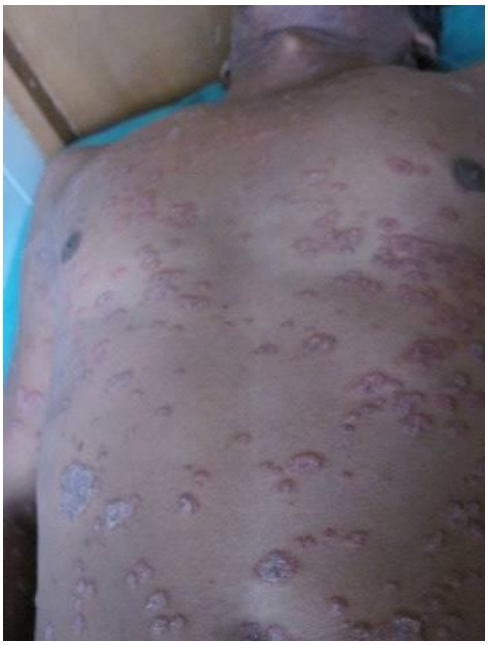
Instep Involvement in Soles



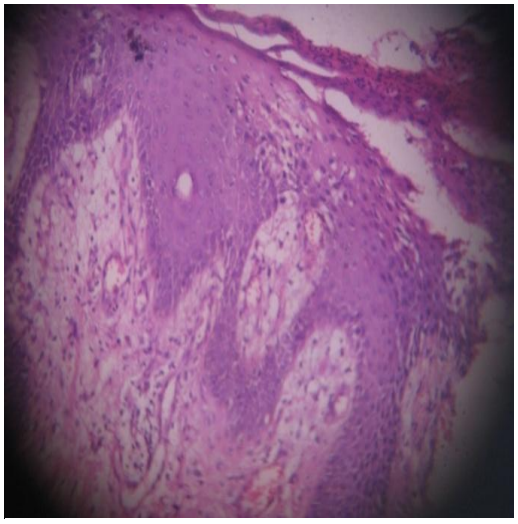
Histopathology

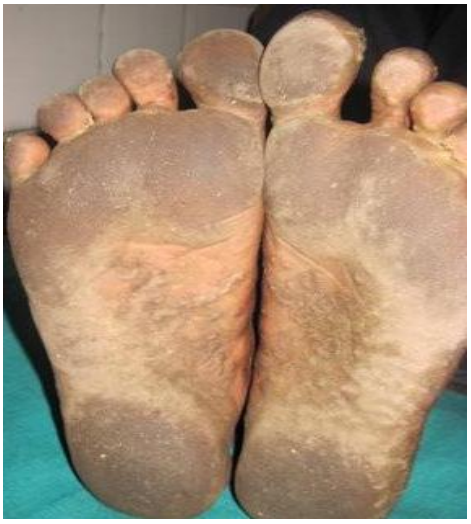
REITERS SYNDROME





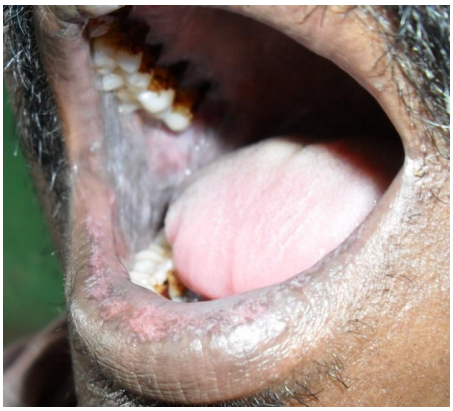
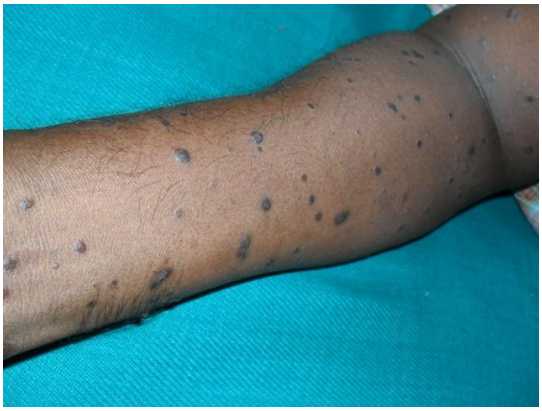
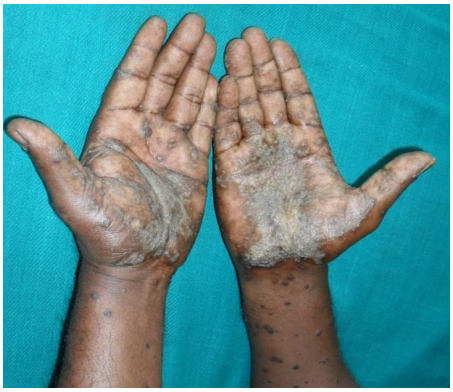
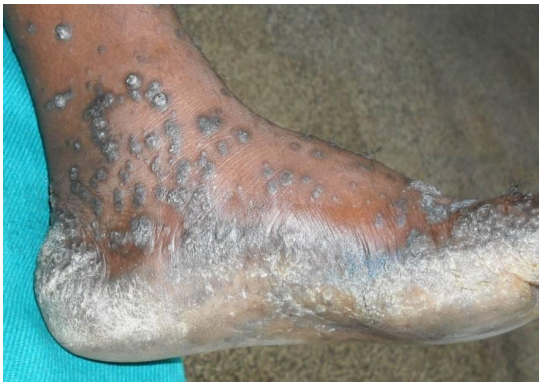
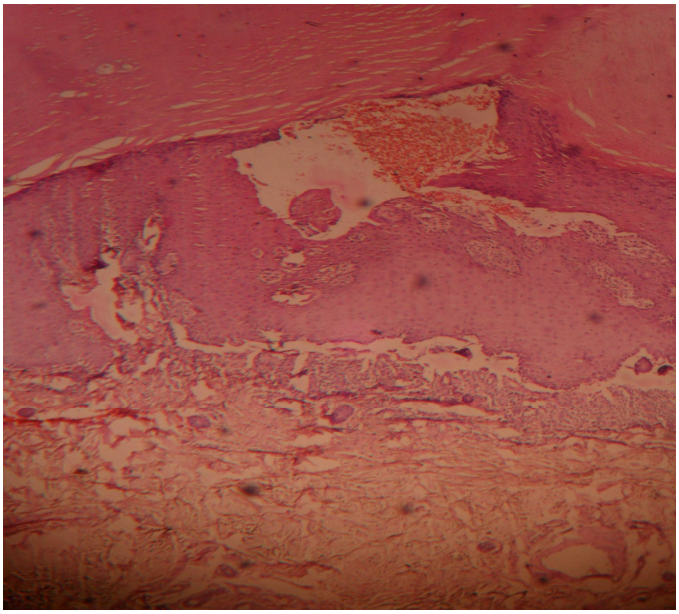
Keratoderma Blenorrhagica





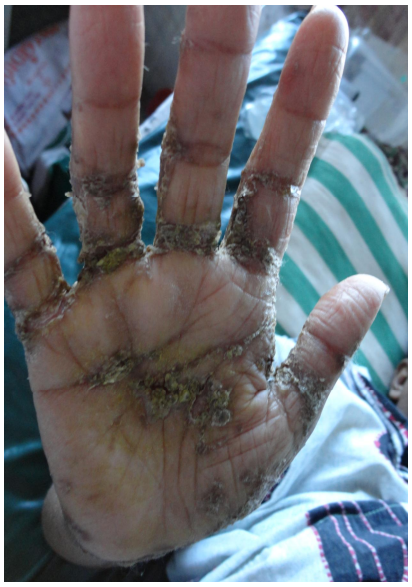
Histopathology

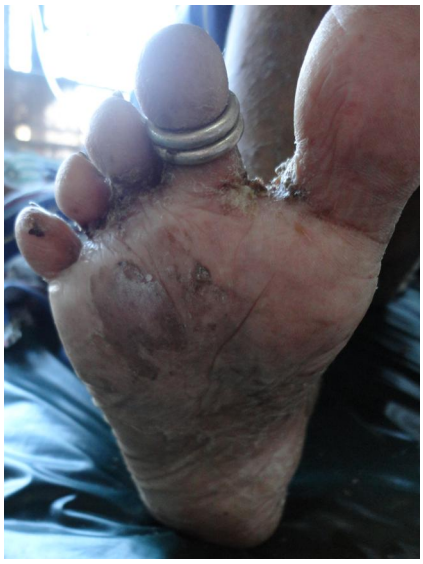
GENERALISED LICHEN PLANUS WITH KERATODERMA

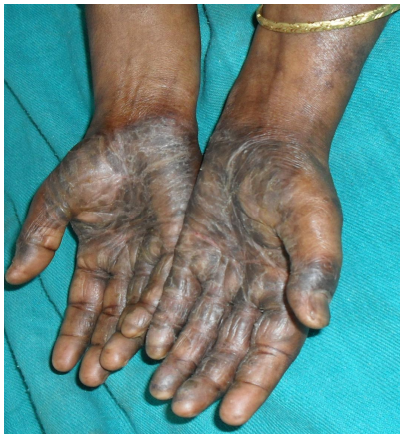
Histopathology

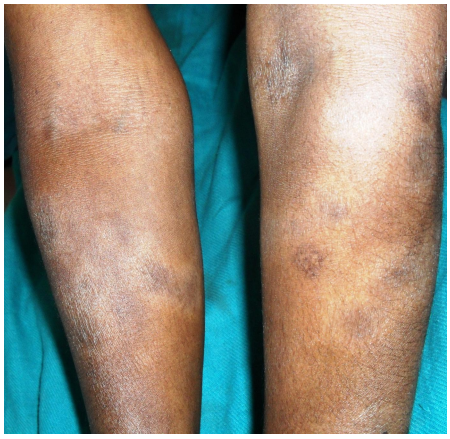
CRUSTED SCABIES

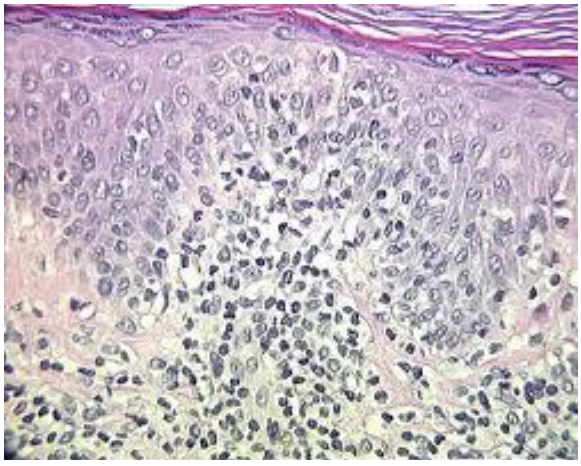




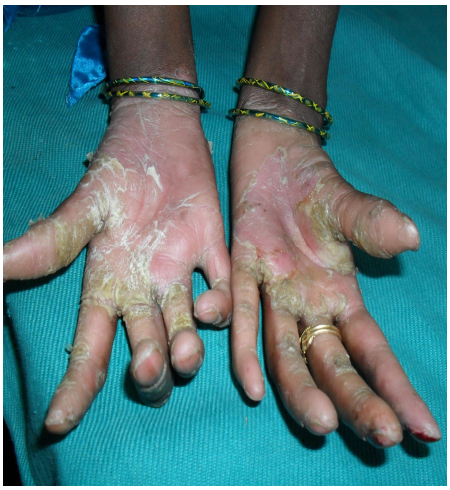
Mycosis Fungoides



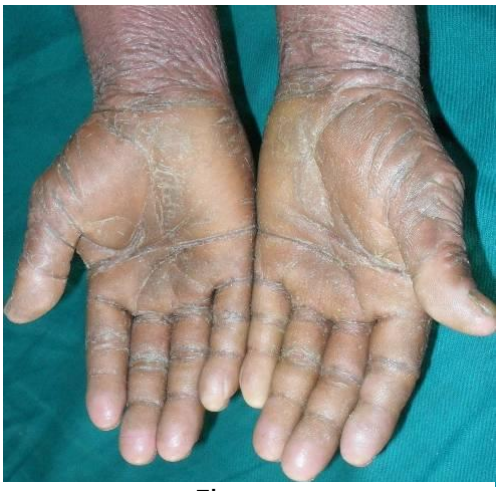




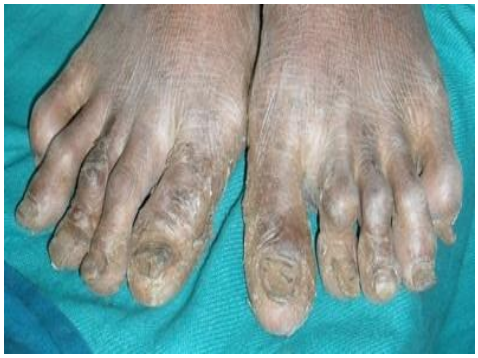
Histopathology



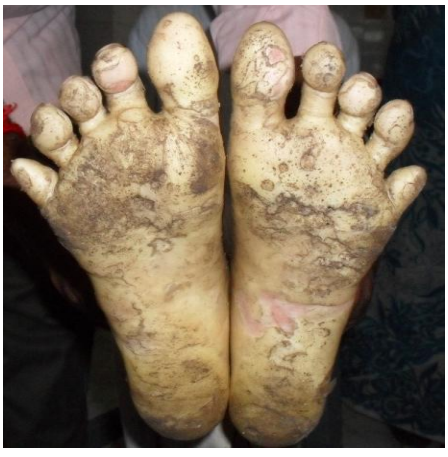
Contractures



Fissures



Nail Changes



Pitted Keratolysis

DISCUSSION

In this study on PPK, with 129 patients the prevalence in our hospital was found to be 0.15%. Out of which hereditary and acquired PPK constituted 0.08% and 0.06% respectively. While Gulati.S, Thappa et al reported the prevalence of hereditary PPK as 5.2 /10,000[11] from Pondicherry. S.C. Murthy et al reported the prevalence as 0.28/100 from Karnataka[10]. Samantal et al from North East India reported the prevalence 0.47 / 100 cases.[105] This shows varied prevalence all over the nation.

Analysis on age incidence revealed that only hereditary PPK was seen during the first decade of life and not acquired PPK. Hereditary PPK was common in the second decade with a prevalence of 37.14%, followed by third decade (28.57%) and first decade(22.85%) and decreased in frequency subsequently in other decades. It was not seen after the sixth decade. Youngest age in which PPK was seen in our study, 2 years in a child with Olmsted Syndrome.

Acquired PPK was commonly seen in the fifth decade (33.89%), followed by the second decade (15.25%). Overall PPK was most

common in the second decade (32.8%). This complies with other study by Mahajan et al who reported 48.76% in the age of 0-15years[12]. Thappa et al reported the highest incidence in the first decade in 67.7% and the average age reported by S.C Murthy et al is 34.53.

Although the age of onset of PPK was since childhood, in this study it was noted during the second decade. This may be due to the fact that limitations of hand movements are most percieved during that time.

Gender wise analysis showed that PPK was more common in males (54.29%) than females(45.73%). In hereditary PPK also males topped the list with 61.42%. However in acquired PPK female preponderance is seen with 54.23%. Mahajan et al also found it to be more common in males. Gulati.S, Thappa et al also reported the male predilection with male to female ratio of 4.2:1. S.C. Murthy et al reported male predominance in acquired PPK also.

In Hereditary PPK unemployed people constituted 18.57%. People remain unemployed probably due to the disease per se, complications and social factors.

In acquired PPK, house wives and manual laborers topped the list with 33.89%,whereas Mahajan et al reported in his study that manual laborer contributed 48.16%, students 33.15% and housewives 18.69%. The majority of the patients were farmers and manual laborers in S.C. Murthy's study. It is surprising to note that some of the people in our study with severe disease were doing jobs which needed fine movements of fingers.

Diffuse nontrangrediens type is the common pattern (77%), among males (47%). Diffuse trangrediens type constituted 21%, of which females were the most common sufferers (14.2%). We have seen only 6 cases of focal type were seen in this study, mainly of focal plantar keratoderma. In the punctate type, only one case was seen during the study period that was a case of marginal keratoderma.

In the acquired PPK focal type was the common pattern (54.27%),seen more commonly among males (28.87%). Diffuse type is less commonly seen (45.7%). Punctuate type of acquired PPK was not seen during the study period. Mahajan et al reported diffuse PPK in 65.21% hereditary PPK as seen in this study. Whereas focal type is the

commonest pattern in S.C. Murthy's study, since majoritiy of their patients belong to acquired PPK, which also supports our study.

Thappa et al reported the incidence of Unna Thost Syndrome as 38.7%. In our study also Unna Thost Syndrome topped the listed with 21 cases (30%). This was followed by focal plantar keratoderma(8.7%). Other syndromes which we have seen are Mal de Meleda, Griethers disease, Olmsted syndrome, Papillon Lefevere syndrome and Naxos disease. Unna Thost Syndrome, was commonly seen in males in this study. Relationship to consanguinity was depicted in table(11). In the Unna Thost syndrome, males(15) outnumbered the females(6).

In those associated with other genodermatosis, ichthyosis vulgaris and familial PRP constituted about 8.57%, Bullous Icthyosiform Erythroderma and Lamellar ichthyosis 4.27%. Other Syndromes seen were kindler syndrome with PPK, Epidermolysis bullosa junctional variety with PPK and Pachydermoperiostitis with PPK were also seen. There was one case of PPK associated with Dowling Degos disease which has not been reported in the literature so far.

Analysis of hereditary PPK had significant correlation with positive family history and offsprings of consanguinous marriage (52.85%) had positive family history of similar illness. 47.14% and 15.71% were offsprings of $3^0$and 2^0 Consanguinity.37.14% belonged to non consanguinous marriage.

Psoriasis constituted about 64.4% of acquired PPK. This include palmoplantar psoriasis and erythrodermic psoriasis with PPK. It include more than 2/3rd in acquired PPK and 1/3 rd of total PPK. This is supported by S.C. Murthy et al and chopra et al[108]. Among paients with psoriasis, females (37.28%) outnumbered the males(27.11%) and was commonly noted in fifth decade. Hyperkeratotic hand and foot eczema constituted about 13%. In this study PPK was seen associated with PRP, crusted scabies, erythroderma, mycosis fungoides, and lichen planus. One case of Keratoderma blenorrhagica in Reiters syndrome was also noted. The case was positive for HLA B27 also. One case of Atopic dermatitis presented with diffuse PPK.

Nail changes associated with PPK, was found to be more in males than females. Pitting was the_predominant finding. Pitted keratolysis was seen in 9 patients. This may be due to hyperhidrosis or wet work

related. Patients with hereditary PPK developed complications like contracture, fissures, bleeding and pain. Systemic association was found to be rare.

Since 18.57% of people in hereditary PPK were unemployed due to disease, an attempt was made to analyze the impact of PPK on physical, social and psychological functions using DLQI[104] measures. It was noted that 22.85% had extremely large effect on life and 47.14% had very large effect on life. In acquired PPK, 64.4% had very large effect on life. But the measures improved with treatment in acquired PPK.

CONCLUSION

- The prevalence of PPK at the Department of Dermatology and Leprosy, Rajiv Gandhi Government General hospital, Madras Medical College, Chennai was 0.15%. Hereditary PPK constituted about 0.08% and acquired PPK constituted 0.06%. In hereditary PPK itself, PPK syndromes and PPK associated with genodermatosis constituted about 61.43% and 38.57% respectively.

- Hereditary PPK was commonly detected in second decade (37.14%). Acquired PPK was commonly seen in the fifth decade (33.89%). Overall it was commonly seen in the second decade 32.8%.

- Gender wise analysis showed that Hereditary PPK was more common in males (54.29%) than females (45.73%). In hereditary PPK a males topped the list with 61.42%. However in acquired PPK female preponderance is seen with 54.23%.

- Acquired PPK was commonly noted in house wives and manual laborers (33.89%).

- Diffuse nontransgrediens type was the common pattern (77%) in hereditary type. Focal type was the common pattern (54.27%) in the acquired type.

- Unna Thost syndrome was the common one. Among which males were commonly affected.

- Others syndromes which was seen in decreasing order of their prevalence were Mal de Meleda, Naxos disease, Griethers disease, Olmsted syndrome, Papillon Lefevere syndrome, Clouston syndrome, Focal plantar keratoderma and Marginal keratoderma.

- Among the PPK associated with other genodermatosis, Ichthyosis Vulgaris was frequently seen followed by Bullous Icthyosiform Erythroderma, Lamellar ichthyosis, Sjogren Larsson syndrome, Familial pitryasis rubra pilaris, Kindler syndrome, Epidermolysis bullosa, Symmetrical progressive erythrokeratoderma, Pachy dermo periosteitis and Dowling Degos disease.

- In the acquired PPK, psoriasis was the common cause followed by hand and foot eczema.

- Psoriasis was common in females and in fifth decade.

- Hereditary PPK had poor DLQI SCORE.

- The common complications was fissuring. Pitted keratolysis is the common secondary infection.

- Systemic associations were found to be rare.

Annexures

REFERENCES

1) R. Judge, W.H.I.Clean, C.S.Munro, Disorders of Keratinization, Rook's Text Book of Dermatology 8th Edition Volume 1.

2) David .P Kelsell, Lrene M. Leigh, Inherited Keratoderma of Palms and Soles. Fitzpatricks Dermatology in general medicine 7th edition. volume 1.

3) Alfrons L Krol Keratodermas,. Dermatology 2nd edition by Jean L Bolognia, Joseph L Jorizzo, Ronald P Rapini.

4) Bernett L,Johnson, Jr. Albert C. Yan. Genodermatoses Lever's histopathology of skin, 10th edition.

5) Arnold Jr HL, Odom RB, James WD. Andrews' diseases of the skin: clinical dermatology. 8th ed. Philadelphia (PA): WB Saunders, 1990.

6) Shaily Patel, Matthew Zirwas, Joseph C Acquired Palmoplantar Keratoderma Am J Clin Dermatol 2007; 8 (1): 1-11 review article 1175-0561/07.

7) Weedon D. Skin pathology. 2nd edition. New York: Churchill Livingstone, 2002.

8) Stevens HP, Kelsell DP, Bryant SP, et al. Linkage of an American pedigree with palmoplantar keratoderma and malignancy (palmoplantar ectodermal dysplasia type III) to 17q24: literature survey and proposed updated classification of the keratodermas. Arch Dermatol 1996; 132: 640-51.

9) Itin PH, Fistrol SK. Palmoplantar keratodermas. Clin Dermatol 2005; 23: 15-22.

10) Sambasiviah Chidambara Murthy.MD,Tanjore Y. Raghu. MD, Tamraparini Suresh .M.D Study of palmoplantar keratodermas in South India. SC, Raghu TY, Suresh T. Int J Dermatol. 2008 Jul;47(7):762-4.

11) Gulati.S. Thappa, Hereditary palmoplantar keratodermas in South India. The Journal of Dermatology [1997, 24(12):765-8].

12) P.M. Mahajan, Vinay Kulkarni Clinical and histopathological study of palmoplantar keratoderma IJDVL 1994 Volume : 60 Issue : 5 Page : 258-26.

13) Requena L, Schoendorff C, Sanchez Yus E Hereditary epidermolytic palmoplantar keratoderma (Vörner type)--report of a family and review of the literature. Clin Exp Dermatol. 1991 Sep;16(5):383-8.

14) Hereditary Palmoplantar (Epidermolytic) Keratoderma: Are Unna-Thost and Vorner the Same Entities?.

15) Hamm H, Happle R, Butterfass T *et al.* Epidermolytic palmoplantar keratoderma of Vörner: is it the most frequent type of hereditary palmoplantar keratoderma *Dermatologica* 1988; **177**: 138–45.

16) Mittal RR, Jha A, Herediatry punctate palmoplantar keratoderma - A clinical study IJDVL Year : 2003 Volume : 69 Issue 2 Page : 90-91.

17) Kimyai-Asadi A, Kotcher LB, Jih MH.Ronald O. The molecular basis of hereditary palmoplantar keratodermas .J Am Acad Dermatol 2002 Sep;47(3):327-43.

18) Reis A,Hennies HC et al Keartin 9 mutations in epidermolytic PPK. Nat Genetics 1994 174-9.

19) Shimizu H Division of Dermatology, Kitasato, Abnormal cornified cell envelope formation in mutilating palmoplantar keratoderma unrelated to epidermal differentiation complex..Journal of investigative dermatology.

20) Ratnavel RC. Grifith WAD the inherited PPK BJD 1997 135. 485-90.

21) Fischer.J,Bouadjar B, Heling Ret al . mutations in gene encoding SLURP -1 In mal de ,meleda . mol.genetics 2001 875-80.

22) Gach JE,Munro cs Lane eb al .two families with Grithers syndrome caused by K1 mutation. JAAD 2005 s 225-30.

23) Fluckiger R, Itin PH. Keratosis extremitatum (Greither's disease): clinical features,histology, ultrastructure. *Dermatology* 1993; **187**: 309–11.

24) Kitasato Grilli R, Aguilar A, Escalonilla P, Soriano L, Fariña C, Martín L, Requena L.Transgrediens et progrediens palmoplantar keratoderma (Greither's disease) with particular histopathologic findings. Cutis. 2000 Mar;65(3):141-5.

25) Aimee L Leonard MD, and Irwin M Freedberg M. Palmoplantar keratoderma of Sybert . Dermatology Online Journal 9(4): 30.

26) Sybert VP, Dale BA, Holbrook KA. Palmar–plantar keratoderma: a clinical, ultrastructural, and biochemical study. *J Am Acad Dermatol* 1988; **18**: 75–86.

27) TS Rajashekar, Gurcharan Singh, Chandra L Naik, Rajendra Okade Camisa disease: A rare variant of Vohwinkel's syndrome IJDVL Year : 2008 Volume : 74 Issue : 1 Page : 81.

28) Camisa C, Rossano C. Variant of keratoderma hereditaria mutilans (Vohwinkel's syndrome). Treatment with orally administered isotretinoin. Arch Dermatol 1984;120:1323-8.

29) Bart RS,Pumphery RE ,Knuckle pads, leuconychia, deafness, a dominantly inhereited syndrome Eng J Medicine 1967 202-7.

30) Richard G et al expanding the phenotype of Cx 26 mutation .Bart-Pumphery syndrome caused by GJB mutation ,J investigative dermatology 2004;128,856-63.

31) keratoderma with scleroatrophy of the the extremities (Huriez syndrome) Delaporte et al BJD 1995 ,133,409-16.

32) Huriez syndrome Shanmugam, sekar, C.R.Srinivas IJDVL 2008 JULY-AUG, VOL 74.

33) Greither's disease S.B. Athanikar ,Arun C. Inamadar , Aparna Palit, V.V. Sampagavi, N.S.Deshmukh. IJDVL 2003 August.vol.69,292-293.

34) Charfeddine C, Mokni M, Ben Mousli R, Elkares R, Bouchlaka C, Boubaker S, Ghedamsi S, Baccouche D, Ben Osman A, Dellagi K, Abdelhak SA novel missense mutation in the gene encoding SLURP-1 in patients with Mal de Meleda from northern Tunisia. Br J Dermatol. 2003 Dec;149(6):1108-15.

35) Nadiga Rajashekhar. Rafeeq Moideen Palmoplantar keratoderma – Mal de Meleda syndrome Int j derm 1997 issue 11 854-856.

36) Mal de Meleda: A report of two cases in one family A. Prohić, E. Kasumagić-Halilović, M.Kantor15.

37) Mozzillo N, Nunziata CA, Caraco C *et al*. Malignant melanoma developing in an area of hereditary palmoplantar keratoderma (Mal de Meleda). *J Surg Oncol*2003; **84**: 229–33.

38) Mahajan VK, Thakur NS, Sharma NL.Papillon-Lefevre syndrome. Indian Pediatr 2003;40:1197-200.

39) Rathi SK. Papillon-Lefevre Syndrome in two brothers. Indian J Dermatol Venerol Leprol 2002;68:155-6.

40) Yagmur A, Yilmaz G, Ertan U, IKizoglu E, Ozkasap S, Karacan C.Papillon Lefevre syndrome: A case report. Int Pediatr 2004;19:224-5.

41) Janjua SA, Khachemoune A.Papillon-Lefevre syndrome: Case report and review of the literature. Dermatol Online J 2004; 10:13.

42) Jigna Shah, Shweta Goe Papillon-Lefevre syndrome: Two case reports IJDVL 2007 Volume : 18 Issue : 4 Page : 210-213.

43) Shahbaz A Janjua , Amor Khachemoune Papillon-Lefèvre syndrome: Case report and review of the literature Dermatology Online Journal 10 (1): 13.

44) Al-Khenaizan S. Papillon-Lefevre syndrome: the response to Acitretin. Int J Dermatol 2002 Dec;41(12):938-41.

45) Haim S, Munk J. Keratosis palmo-plantaris congenita, with periodontosis, arachnodactyly and a peculiar deformity of the terminal phalanges. *Br J Dermatol* 1965;

46) Clouston HR. A hereditary ectodermal dystrophy. *Can Med Assoc J* 1921; **21** 18– 31.

47) Maurice A M van Steensel, Marcel F Jonkman*, Michel van Geel, Peter M Steijlen, W H Irwin McLean† and Frances J D Smith†Clouston Syndrome Can Mimic Pachyonychia Congenita Journal of Investigative Dermatology (2003) 121, 1035–1038.

48) Pramod Kumar, PK Sharma, HK Kar Olmsted syndrome Indian Journal of Dermatology, Year 2008, Volume 53, Issue 2 [p. 93-95].

49) Dessureault J et al.Olmsted syndrome associated with malignant melanomas. j cutan med surg 2003 236-42.

50) Vijaikumar M, Thappa DM, Jeevankumar B.An Indian case of keratoderma hereditarium mutilans (Vohwinkel's syndrome) associated with ichthyosiform dermatosis. J Dermatol. 2001 Oct;28(10):560-3.

51) Perish .k , Salvati .EF. Keratoderma hereditarium mutilans (Vohwinkel's syndrome) associated with congenital deaf-mutism. Br J Dermatol. 1995 Apr;132(4):617-20.

52) Meravu .B, Golberg .g et al Olmsted syndrome: mutilating palmoplantar keratoderma with periorificial keratotic plaques. J Am Acad Dermatol. 2005 Nov;53.

53) Berad MS.Powell.J et al ,Palmoplantar keratoderma and skin grafting: postsurgical long-term follow-up of two cases with Olmsted syndrome. Pediatr Dermatol. 2008 Mar-Apr;25(2):223-9.

54) R Rai1, B Ramachandran1, VS Sundaram1, G Rajendren2, CR Srinivas Naxos disease: A rare occurrence of cardiomyopathy with woolly hair and palmoplantar keratoderma IJDVL Year: 2008 , Volume : 74, Issue : 1, Page : 50-52.

55) Protonotarios N, Tsatsopoulou A, Patsourakos P, Alexopoulos D, Gezerlis P, Simitsis S, et al. Cardiac abnormalities in familial palmoplantar keratosis. Br Heart J 1986;56:321-6.

56) Carvajal-Huerta L. Epidermolytic palmoplantar keratoderma with wooly hair and dilated cardiomyopathy. J Am Acad Dermatol 1998;39:418-21.

57) Wachters DHJ, Frensdorf EL, Hausman R *et al*. Keratosis palmoplantaris nummularis ((‘hereditary painful callosities’). *J Am Acad Dermatol* 1983; **9**: 204–9.

58) Howel-Evans W, McGonnell RB, Clarke GA, Sheppard PM. Carcinoma of the oesophagus with keratosis palmaris et plantaris (tylosis): a study of two families. *QJM* 1950; **27**: 415–29.

59) RPatnaik, M Krishnaswamy ,Keratosis Palmaris Et Plantaris (tylosis) with Carcinoma Oesophagus IJDVL 1963 Volume : 29 Issue : 6 Page : 225—227.

60) Lalitha Janakiraman, Malathi Sathiyasekaran, Munirathiram Deenadayalan, Ramaswamy Ganesh and Uma Mahesh Richner Hanhart Syndrome Indian J Pediatr 2006; 73 (2) : 161-162.

61) Viglizzo GM, Occella C, Bleidl D, Rongioletti F. Richner–Hanhart syndrome (tyrosinemia II): early diagnosis of an incomplete presentation with unusual fi ndings. *Pediatr Dermatol* 2006; **23**: 259.

62) Su WP, Chun SI, Hammond DE, Gordon H. Pachyonychia congenita : a clinical study of 12 cases and review of the literature. Pediatar Dermatol 1990; 7 : 33-8.

63) Feinstein A, Friedman J, Schewach-Millet M. Pachyonychia congenita. J Am Acad Dermatol 1988; 19 : 705-11.

64) A K Jaiswal, S Badrinath, J Pillai, M Bhardwaj, Pachyonychia congenita tarda IJDVL 1994 vol 60 page43-44.

65) Paller SA, Moore JA, Seher R. Pachyonychia congenita tarda : A late-onset form of Pachyonychia congenita. Arch Dermatol 1991; 127 : 701-3.

66) Marco Castori, Schöpf-Schulz-Passarge Syndrome: Further Delineation of the Phenotype and Genetic Considerations Acta Derm Venereol 2008; 88: 607–612.

67) Hampton PJ, Angus B, Carmichael AJ. A case of Schopf–Schulz–Passarge syndrome. *Clin Exp Dermatol* 2005; **30**: 528–30.

68) Marco Castori, Brauer-Buschke-Fischer keratoderma palmo-plantare punctata. Brauer's keratoderma dissipatum hereditarium palmoplantare ini Dermatologia, 1990 Nov;125(11):527-32.

69) Jose S, Kamath KN, Pai GS, Pinto J Punctate palmoplantar keratoderma IJDVL 2003 Volume : 69 Issue : 7 Page : 70-71.

70) Hesse S, Berbis PH, Privat Y. Keratoderma palmoplantaris papulosa (Buschke-Fischer's disease). Efficacy of actitretin Br J Dermatol 1993; 128:104-105.

71) Emmert S, Kuster W, Hennies HC *et al.* 47 patients in 14 families with the rare genodermatosis keratosis punctata palmoplantaris Buschke-Fischer-Brauer. *Eur J Dermatol* 2003; **13**: 16–20.

72) Powell FC, Venencie PY, Gordon H et al. Keratoderma and spastic paralysis. Br J Dermatol 1983; 109:29-34.

73) Renu Roy, G Kalla, Asheesh Batra Acrokerato elastoidosis of costa: Report of three cases Indian Journal of Dermatology, Venereology, and Leprology, Year 1997, Volume 63, Issue 5 [p. 312-313].

74) Deschamps P, Leroy D, Pedailles S, et al.Haxthausen H. Keratoderma climactericum. Br J Dermatol 1934; 46: 161-7.

75) Keratoderma climactericum (Haxthausen's disease): clinical signs, laboratory findings and etretinate treatment in patients. Dermatologica 1986; 172: 258-62.

76) Wathcel TJ. Plantar and palmar hyperkeratosis in young castrated women. Int J Dermatol 1981; 20: 270-1.

77) Brincat M, Moniz CJ, Studd JW, et al. Long term effects of the menopause and sex hormones on skin thickness. Br J Obstet Gynaecol 1985; 92: 256-9.

78) Shuster S, Black MM, McVitie E. The influence of age and sex on skin thickness, skin collagen and density. Br J Dermatol 1975; 93: 639-43.

79) Zultak M, Bedeaux C, Blanc D. Keratoderma climactericum treatment with topical - estrogen. Dermatologica 1988; 176: 151-2.

80) Sass U, Grosshans E, Simonart JM. Chronic arsenicism: criminal poisoning or a drug-intoxication? Dermatology 1993; 186: 303-5.

81) Poskitt LB, Duffill MB, Rademaker MR. Chloracne, palmoplantar keratoderma and localized scleroderma in a weed sprayer. Clin Exp Dermatol 1994; 19: 264.

82) Shannon RS, Strayer DS. Arsenic induced skin toxicity. Hum Toxicol 1989; 8: 99.

83) Rahman MM, Chowdhury UK, Mukherjee SC, et al. Chronic arsenic toxicity in Bangladesh and West Bengal, India: a review and commentary. J Toxicol Clin Toxicol 2001; 39 (7): 683-700.

84) Tindall JP. Chloracne and chloracnegens. J Am Acad Dermatol 1985; 13: 539-58.

85) Geusau A, Jurecka W, Nahavandi H, et al. Punctate keratoderma-like lesions on the palms and soles in a patient with chloracne: a new clinical manifestation of dioxin intoxication? Br J Dermatol 2000; 143 (5): 1067-71.

86) Lee BY, Hogan DJ, Ursine S, et al. Personal observation of skin disorders in malnutrition. Clin Dermatol 2006; 24: 222-7.

87) Duvic M, Reisman M, Finley V, et al. Glucan-induced keratoderma in acquired immunodeficiency syndrome. Arch Dermatol 1987; 123: 751-6.

88) Albersheim P, Darvill AG. Oligosaccharins. Sci Am 1985; 253: 58-64 Jucgla A, Sais G, Navarro M, et al. Palmoplantar keratoderma secondary to chronic acral erythema due to tegafur. Arch Dermatol 1995; 131 (3): 364-5.

89) A case of syphilitic keratoderma concurrent with syphilitic uveitis.Ann Dermatol. 2009 Nov;21(4):399.

90) Munro CS, Cox NH, Marks JM, et al. Lichen nitidus presenting as palmoplantar hyperkeratosis and nail dystrophy. Clin Exp Dermatol 1993; 18: 381-3.

91) Kumar B, Saraswat A, Inderjeet K. Palmoplantar lesions in psoriasis: A study of 3065 patients. Acta Derm Venereol 2002; 82: 192-5.

92) Yan AC, Aasi SZ, Alms WJ, et al. Aquagenic palmoplantar keratoderma. J Am Acad Dermatol 2001; 44: 696-9.

93) Neri I, Bianchi F, Patrizi A. Transient aquagenic palmar hyperwrinkling: the first instance reported in a young boy. Pediatr Dermatol 2006; 23 (1): 39-42.

94) Herman PS. Punctate porokeratotic keratoderma. Dermatologica 1973; 147: 206-13.

95) Rault S, Salmon Ehr V, Cambie MP, et al. Palmoplantar filiform parakeratotic hyperkeratosis and digestive adenocarcinoma. Ann Dermatol Venereol 1997 124 (10): 707-9.

96) Miller JJ, Roling D, Spiers E, et al. Palmoplantar keratoderma associated with hypothyroidism. Br J Dermatol 1998; 139: 741-2.

97) Hodak E, David M, Feuerman EJ. Palmoplantar keratoderma in association with myxedema. Acta Derm Venerol 1986; 66: 354-7.

98) Bazex A, Griffiths A. Acrokeratosis paraneoplastica: a new cutaneous marker of malignancy. Br J Dermatol 1980; 102: 301.

99) Jacobsen FK, Abildtrup N, Laursen SO, et al. Acrokeratosis paraneoplastica (Bazex' syndrome). Ach Dermatol 1984; 120: 502-4.

100) Lo WL, Wong CK. Tripe palms: a significant cutaneous sign of internal malignancy. Dermatology 1992; 185: 151-3.

101) Sehgal VN, Wagh SA. Cutaneous tuberculosis : current concepts. Int J Dermatol 1990; 29: 237-252.

102) Pramatarov K, Balbanova M, Miteva L, et al. Tuberculosis verrucosa cutis associated with lupus vulgaris. Int. J Dermatol 1993; 32:815-817.

103) Lee BY, Hogan DJ, Ursine S, et al. Personal observation of skin disorders in malnutrition. Clin Dermatol 2006; 24: 222-7.

104) Finlay AY, Khan GK. Dermatology Life Quality Index (DLQI): a simple practical measure for routine clinical use. *Clin Exp Dermatol* 1994; **19**: 210–6.

105) Samanta BC, Banerjee BN, Panja RK. Etiology of plantar keratoderma. *Indian J Dermatol Venereol Leprol* 1976; **42**:116–125.

106) Chopra A, Maninder Gill SS. Hyperkeratosis of palms and soles: clinical study. *Indian J Dermatol Venereol Leprol* 1997; **63**: 85–88.

107) Alsaleh QA, Teebi AS. Autosomal recessive epidermolytic palmoplantar keratoderma. *J Med Genet.*1990;27:519-522.

108) Nogita T, Nakagawa H, Ishibashi Y. Hereditary epidermolytic palmoplantar keratoderma with knuckle pad-like lesions over the finger joints. *Brit J Dermatol.* 1991;125:496.

109) Barraud-Klenovsek MM, Lubbe J, Burg G. Primary digital clubbing associated with palmoplantar keratoderma. *Dermatology.* 1997;194:302-305.

110) Miteva L, Dourmishev AL. Hereditary palmoplantar keratoderma with nail changes and periodontosis in a father and son. *Cutis.* 1999;63:65-68.

111) Bennion SD, Patterson JW. Keratosis punctata palmaris et plantaris and adenocarcinoma of the colon. A possible familial association of punctate keratoderma and gastrointestinal malignancy. *J Am Acad Dermatol.* 1984;10:587-591.

112) Camisa C, Hessel A, Rossana C, Parks A. Autosomal dominant keratoderma, ichthyosiform dermatosis and elevated serum β-glucuronidase. *Dermatologica* 1988; **177**: 341–7.

113) Fluckiger R, Itin PH. Keratosis extremitatum (Greither's disease): clinical features,histology, ultrastructure. *Dermatology* 1993; **187**: 309–111.

114) Haim S, Munk J. Keratosis palmo-plantaris congenita, with periodontosis, arachnodactyly**and** a peculiar deformity of the terminal phalanges. *Br J Dermatol* 1965;**77**: 42–54.

PROFORMA

Date :

Address :

Case No. :

Name :

Age :

Sex :

Complaints :

HISTORY OF PRESENT ILLNESS

Age of onset of disease

H/o Involvement beyond the dorsal hand and feet

H/o Discolouration – redness / yellow

H/o Hyperhidrosis

H/o Hypohidrosis

H/o Blistering

H/o Pyoderma

H/o Itching

H/o Oozing

H/o Constructing bands over digits

H/o Auto amputation of digits

H/o Dryness of skin, seasonal variation

H/o Colloidon baby

H/o Woolly hair

H/o. Hair loss

H/o Nail Involvement

H/o Loss of teeth / teeth anomalies

H/o Hearing defect

H/o Visual defect

H/o Cardiac symptoms – Dyspnea,Paraoxysmal Nocturnal Dyspnea Orthopnea / Palpitation / Giddiness

H/o Dysphagia

History Suggestive of hypothyroidism

History Suggestive of internal malignancy

H/o Arsenical intake

H/o exposure to Sexually transmitted diseases

PAST HISTORY

H/o DM/ HT / PT

HISTORY

Pedigree chart

Consanguinity

Other siblings affected

OCCUPATIONAL HISTORY

GENERAL EXAMINATION

Nourishment

Anaemia

Lymph adenopathy

Jaundice

Mentation

Vision

Hearing

Vital signs

System examination

Cardiovascular system

Respiratory system

Abdomen

Central Nervous System

DERMATOLOGICAL EXAMINATION OF PALMS AND SOLES

- Diffuse
- Focal
  - Areata
  - Striate
- Punctate

- Colour
  - Erythema
  - Yellow
- Surface
  - Smooth
  - Verrcous
  - Pitted
- Transgrediens ±
- Pseudo ainhum
- Knuckle pads
- Blisters
- Pyoderma

OTHER SKIN AREAS

NAILS

Leuconychia

Nail drystrophy

Koilonychias

Thickened nail

Sub ungual hyperkeratosis

Fissuring

Ridging of nails

HAIR

Woolly hair

Alopecia

MUCOSA

Oral mucosa

Genital mucosa

TEETH

INVESTIGATION

Complete blood count

LFT

RFT

Lipid profile

STS

Skin biopsy

Scraping for fungal examination and mite

Special investigation if any

Specialty opinion

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
1	14	2	F	H	5	S	-	-	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	14	F
2	11	2	M	H	1	U	-	-	+	-	-	+	-	-	LA	-	+	-	+	UT	-	-	22	-
3	13	2	F	H	2	S	-	3	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	15	F
4	16	2	M	H	5	M	+	2	-	-	-	+	-	-	-	-	-	-	+	PLS	-	-	21	-
5	29	3	M	H	10	L	+	3	+	-	-	-	-	-	KP	-	-	-	-	UT	-	-	20	F,B
6	24	3	M	H	10	O	+	3	+	-	-	-	-	-	KP	-	-	-	-	-	IV	-	12	-
7	27	3	M	H	5	P	+	2	-	-	-	+	+	+	KP	-	+	-	-	OMS	-	-	24	C,PA,AD
8	10	2	F	H	6	S	-	-	+	-	-	+	-	-	-	-	-	-	-	UT	-	-	8	-
9	6	1	M	H	5	S	-	-	-	+	-	-	-	-	-	-	-	-	-	FPK	-	-	7	-
10	13	2	M	H	13	S	-	-	-	+	-	-	-	-	-	-	+	-	+	FPK	-	-	9	F
11	19	2	F	A	15	U	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	7	F,B
12	40	5	M	A	40	L	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PRP	12	-
13	20	3	M	H	2	S	+	-	-	-	+	-	-	-	-	-	-	-	-	MG	-	-	9	-
14	14	2	F	H	6	S	-	-	+	-	-	-	-	-	KP	-	-	-	-	-	FPRP	-	8	F
15	40	5	M	A	39	L	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	ECZ	15	-
16	35	4	M	A	35	L	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	ERD	20	F,B
17	65	7	F	A	65	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	7	F
18	44	5	M	A	44	L	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	14	-
19	18	2	F	A	17	S	-	-	+	-	-	-	-	-	-	+	+	-	-	-	-	PSOR	17	E,F
20	49	5	F	A	49	L	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	15	F
21	50	6	F	A	49	H	+	3	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	10	-
22	13	2	F	H	1	U	-	-	+	-	-	+	+	+	KP	+	+	+	+	OMS	-	-	25	C,F,P,B

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
23	46	5	F	A	35	L	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	AD	14	C
24	35	4	M	A	34	F	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	RTR	17	-
25	20	3	M	H	1	L	-	2	+	-	-	-	-	-	-	-	-	-	+	UT	-	-	14	C,F
26	82	9	F	A	81	U	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ERD	16	F
27	70	8	F	A	70	H	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	PSOR	13	-
28	40	5	F	A	39	H	-	3	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	12	-
29	7	1	M	H	1	S	+	-	+	-	-	-	-	-	-	-	-	-	+	-	IV	-	8	-
30	45	5	F	H	7	H	+	3	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	5	-
31	22	3	M	H	3	E	-	-	+	-	-	-	-	-	-	-	+	-	-	-	IV	-	17	C,SA
32	45	5	M	H	20	F	-	-	+	-	-	-	-	-	-	-	+	+	-	UT	-	-	11	F
33	50	6	F	H	5	H	+	3	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	12	-
34	36	4	M	H	17	F	+	3	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	17	-
35	25	3	M	H	21	L	-	-	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	5	-
36	27	3	M	H	2	L	+	3	+	-	-	-	-	-	KP	-	-	-	-	UT	-	-	19	F
37	12	2	M	H	1	U	+	2	+	-	-	-	-	-	-	-	+	-	+	-	LI	-	24	C,F
38	13	2	M	H	1	U	+	2	+	-	-	-	-	-	-	-	+	-	+	-	LI	-	25	C,F
39	15	2	M	H	1	U	+	3	+	-	-	-	-	-	-	-	-	-	-	-	LI	-	20	F
40	10	2	M	H	5	M	-	3	+	-	-	-	-	-	KP	-	+	-	-	UT	-	-	13	-
41	22	3	M	H	1	W	+	3	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	12	-
42	50	6	F	H	1	H	-	2	+	-	-	+	-	+	KP	-	+	-	+	MM	-	-	19	-
43	29	3	F	H	1	H	+	2	+	-	-	-	-	-	-	+	+	-	+	-	IV	-	16	-
44	28	3	F	H	1	C	+	3	+	-	-	-	-	-	-	-	-	-	-	-	BIE	-	21	C

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
45	25	3	M	H	1	C	+	3	+	-	-	-	-	-	-	-	-	-	-	-	BIE	-	20	-
46	25	3	F	H	1	H	+	3	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	11	-
47	13	2	F	H	2	S	-	-	+	-	-	-	-	-	-	-	-	-	-	-	FPRP	-	5	-
48	26	3	M	H	8	C	-	-	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	18	F
49	12	2	F	H	1	S	-	2	+	-	-	+	+	+	KP	-	+	+	+	MM	-	-	23	C,SA,F
50	11	2	F	H	1	U	+	3	+	-	-	-	+	-	-	+	+	+	-	NX	-	-	22	C
51	8	1	M	H	1	U	+	3	+	-	-	-	-	-	-	-	-	-	-	NX	-	-	23	C
52	18	2	F	H	1	O	+	3	-	-	-	+	+	+	KP	-	+	-	-	MM	-	-	26	C,SA,
53	15	2	F	H	1	S	+	3	-	-	-	+	+	+	KP	-	+	-	-	MM	-	-	22	C
54	27	3	M	H	7	E	-	-	+	-	-	-	-	-	-	-	+	-	-	UT	-	-	18	F
55	2	1	f	H	1	S	+	2	+	-	-	+	+	+	KP	-	+	-	-	OMS	-	-	17	C,
56	35	4	m	H		O	+	3	+	-	-	-	-	-	-	-	-	-	-	-	BIE	-	16	-
57	15	2	f	H	2	U	+	3	+	-	-	+	-	-	-	-	-	-	+	PLS	-	-	19	C
58	22	3	m	H	1	S	-	-	+	-	-	-	-	-	-	-	+	-	-	-	KND	-	16	I
59	13	2	F	H	1	U	-	2	+	-	-	-	-	-	-	-	-	-	+	-	I-SLS	-	23	-
60	8	1	F	H	1	S	-	3	+	-	-	+	+	-	KP	+	+	-	-	CL	-	-	21	C
61	45	5	F	A	45	H	+	-	+	-	-	-	-	-	-	-	-	-	+	-	-	CS	13	-
62	22	3	F	A	22	U	-	-	-	+	-	-	-	-	-	-	-	-	-	-	-	CS	25	-
63	8	1	M	H	2	S	+	3	+	-	-	-	-	-	-	-	-	-	-	-	IV	-	7	-
64	55	6	F	A	54	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	11	-
65	8	1	M	H	2	S	+	-	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	13	F
66	10	2	M	H	5	S	-	-	-	+	-	-	-	-	-	-	-	-	-	FPK	-	-	4	F

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
67	9	1	M	H	4	S	+	3	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	10	-
68	2	1	M	H	1	S	-	-	-	+	-	-	-	-	-	-	-	-	-	FPK	-	-	4	-
69	50	6	M	A	50	C	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PRP	18	F,B
70	8	1	F	H	1	S	+	3	+	-	-	-	-	-	-	-	-	-	-	-	EBD	-	15	AD
71	70	8	M	A	70	M	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	11	-
72	19	2	M	H	1	U	+	3	+	-	-	+	+	+	KP	-	+	-	-	GR	-	-	25	C,F,E
73	12	2	M	H	1	S	+	3	+	-	-	+	+	+	-	-	+	-	-	GR	-	-	18	C,F,B
74	6	1	F	H	1	S	-	-	+	-	-	+	+	+	PD	-	-	-	-	GR	-	-	20	C
75	3	1	M	H	1	S	+	3	+	-	-	-	-	-	-	-	-	-	-	-	IV	-	7	-
76	23	3	F	H	15	H	-	3	+	-	-	-	-	-	PD	-	+	-	-	-	FPRP	-	8	-
77	5	1	M	H	4	S	-	-	+	-	-	-	-	-	-	-	-	-	-	-	FPRP	-	5	-
78	13	2	M	H	1	S	-	3	+	-	-	-	-	-	-	-	+	-	-	-	KND	-	14	-
79	15	2	M	H	1	S	-	2	+	-	-	-	-	-	-	-	+	-	-	-	KND	-	16	-
80	24	3	M	H	1	U	+	3	+	-	-	-	-	-	-	-	+	-	-	-	KND	-	22	-
81	39	4	M	A	39	L	-	-	-	+	-	-	-	-	-	-	-	-	-	-	-	PSOR	9	-
82	12	2	M	H	6	S	-	3	+	-	-	-	-	-	-	-	-	-	-	UT	-	-	5	-
83	45	5	M	H	35	L	+	-	-	-	-	-	-	-	-	-	+	-	+	-	PDB	-	16	-
84	24	3	M	H	1	C	-	-	+	-	-	-	-	-	PD	-	+	-	-	UT	-	-	18	F
85	75	8	M	A	75	U	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PRP	19	C,F
86	15	2	M	H	8	S	-	-	+	-	-	-	-	-	-	-	+	-	-	-	FPRP	-	13	-
87	38	4	M	A	35	F	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PRP	21	-
88	8	1	F	H	3	S	+	3	+	-	-	-	-	-	-	-	-	-	-	FPK	-	-	3	F

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
89	4	1	F	H	2	S	+	3	+	-	-	-	-	-	-	-	-	-	-	FPK	-	-	4	F
90	20	3	F	H	8	U	-	-	+	-	-	-	-	-	-	-	+	-	-	-	FPRP	-	8	-
91	40	5	F	H	39	H	+	3	+	-	-	-	-	-	-	-	-	-	-	-	DDD	-	18	-
92	8	1	M	H	1	U	-	-	+	-	-	-	-	-	-	-	-	-	-	-	SPEK	-	6	-
93	45	5	F	A	45	H	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	MF	16	F
94	60	7	F	A	60	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	15	-
95	40	5	F	A	40	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	12	-
96	40	5	M	A	40	M	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	13	F
97	40	5	F	A	40	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	ECZ	11	-
98	27	3	F	A	27	O	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	17	-
99	55	6	M	A	55	L	-	-	-	+	-	-	-	-	-	-	+	+	-	-	-	PSOR	15	-
100	60	7	M	A	60	L	-	-	-	+	-	-	-	-	-	+	+	-	-	-	-	PSOR	16	-
101	45	5	F	A	45	H	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	14	-
102	40	5	M	A	40	L	-	2	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	13	-
103	27	3	F	A	27	H	-	-	-	+	-	-	-	-	-	-	+	+	-	-	-	PSOR	11	-
104	40	5	F	A	40	H	-	-	-	+	-	-	-	-	-	+	+	-	-	-	-	PSOR	9	-
105	45	5	F	A	45	H	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	8	-
106	56	6	M	A	56	O	+	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	11	-
107	42	5	F	A	42	L	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	12	-
108	50	6	F	A	50	H	-	3	-	+	-	-	-	-	-	-	+	+	-	-	-	PSOR	9	-
109	36	4	M	A	36	L	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	13	-
110	28	3	M	A	28	F	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	16	-

Case. No.	Age	Age Decade	Sex	H/A	O.s.D	Occu	F. H	Cons.	D	F	P	T	Eryt	M-o	Sec. Inf.	Hair	Nail	Mucosa	Sys. Assn.	PPK	G.D	APPK	DLQI	Complic
111	29	3	M	A	29	L	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	15	-
112	25	3	M	A	25	L	-	2	-	+	-	-	-	-	-	-	+	+	-	-	-	PSOR	10	-
113	50	6	F	A	50	H	-	-	+	-	-	-	-	-	-	-	-	-	-	-	-	PSOR	8	-
114	50	6	M	A	50	C	-	3	-	+	-	-	-	-	-	+	+	-	-	-	-	PSOR	7	-
115	48	5	M	A	48	L	+	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	6	-
116	30	4	F	A	30	H	-	-	-	+	-	-	-	-	-	-	-	-	-	-	-	PSOR	11	-
117	22	3	F	A	22	U	-	-	-	+	-	-	-	-	-	-	-	-	-	-	-	PSOR	10	-
118	35	4	M	A	35	L	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	15	-
119	43	5	F	A	43	H	-	3	-	+	-	-	-	-	-	-	-	-	-	-	-	PSOR	7	-
120	21	3	F	A	21	S	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	6	-
121	45	5	M	A	45	F	-	-	+	-	-	-	-	-	-	+	+	-	-	-	-	LP	13	-
122	15	2	M	A	15	L	-	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	5	-
123	17	2	M	A	17	S	-	2	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	4	-
124	16	2	M	A	18	S	+	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	6	-
125	19	2	F	A	19	S	+	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	8	-
126	20	3	F	A	20	U	-	3	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	13	-
127	79	8	F	A	79	H	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	KP	16	F
128	45	5	F	A	45	H	+	-	-	+	-	-	-	-	-	-	+	-	-	-	-	PSOR	7	-
129	38	4	M	A	38	L	-	-	+	-	-	-	-	-	-	-	+	-	-	-	-	PSOR	12	-

KEY TO MASTER CHART

M	:	Male
F	:	Female
H	:	Hereditary
A	:	Acquired
A.O.D.	:	Age of onset of Disease
Occ	:	Occupation `S' – Student `U' – Employed `M' – Mechanic, `L' Laborer `O' – Office Work P- Painter `H' Housewife `F' Fisherman `W' Weaver `E' Engineer
F.H	:	Family History
Cons	:	Consanguinity
D	:	Diffuse
F	:	Focal
P	:	Punctate
`T'	:	Transgredient
E.B	:	Erythematous borders
M.O.	:	Malodour
S.I.	:	Secondary Infection, K.P. – Keratolysis Punctata, P.D – Pyoderma. L.A. – Liver Abscess

Sys.Ass	:	Systemic Association A – Anaemia CHD Congenital Heart Disease B.A – Brolchial Asthma, SLE - Systemic Lupus Erythematosis, R.A. – Rheumatoid Arthritis
G.D.	:	Genodermatosis
DLQI	:	Dermatological Life Quality Index
Comp	:	Complications C - Contracture, F- Fissures B - Bleeding, P.A. Pseudo Ainhum E – Erythema
UT	:	Unna-Thost syndrome
MM	:	Mal de Meleda
GD	:	Greither's Disease
CLO		Clouston Syndrome
OLM		Olmsted Syndrome
PLS	:	Papillon-Lefevre Syndrome
NX	:	Naxos Disease
BIE		Bullous Congenital Ichthyosiform Erythroderma,
LI	:	Lamellar Ichthyosis
SLS	:	Sjogren Larsson Syndrome
IV	:	Ichthyosis Vulgaris
FPRP	:	Familial Pityriasis Rubra Pilaris
PRP	:	Pityriasis Rubra Pilaris

SPEK	:	Symmetric Progressive Erythro Keratodermas
AD	:	Atopic Dermatitis
CS	:	Crusted Scabies
ECZ	:	Eczema
PSOR	:	Psoriasis
RD	:	Reiter’s Diseases
ERD	:	Erythroderma
FPK	:	Focal Plantar Keratoderma
M.F	:	Mycosis Fungoides
K.S.	:	Kindler Syndrome
EBS	:	Epidermolysis Bullosa
DDD	:	Dowling Degos Disease
PDP	:	Pachy Dermo Periostetitis

ABBREVIATIONS

PPK	:	Palmo Plantar Keratoderma
EPPK	:	Epidermolytic Palmo Plantar Keratoderma
NEPPK	:	Non Epidermolytic Palmo Plantar Keratoderma
HPPK	:	Hereditary Palmo Plantar Keratoderma
A.D	:	Autosomal Dominant
A.R.	:	Autosomal Recessive
XLR	:	`X' Linked Recessive
TSH	:	Thyroid Stimulating Hormone
CXR	:	Chest X ray
CBC	:	Complete Blood Count
ANA	:	Anti Nuclear Antibody
RPR	:	Rapid Plasma Reagin Test
MX	:	Mantoux
CX	:	Connexin
GJ	:	Gap Junction