# DOWNLOAD PDF CONNECTIVE TISSUE DISEASES OF THE SKIN (CLINICAL DERMATOLOGY)

## Chapter 1 : Connective Tissue Disease of the Skin | JAMA Network Collections | JAMA Network

*Connective Tissue Disease of the Skin Explore the latest in connective tissue disease of the skin, including advances in understanding its diagnosis and management. Add to Your Interests.*

Non-erosive arthritis Myositis and associated weakness Neurological Have you ever had a seizure or an episode of psychosis? If so, was an underlying etiology found? Lupus cerebritis Rare reports of preceding trigeminal neuralgia Hematologic Has anyone ever told you that you had low blood counts? Have you ever required a transfusion? Cytopenias Rare cytopenia Renal Do you have kidney disease? Have you been told you have protein in your urine? Have you ever been hospitalized for kidney problems? Do you know your hepatitis B and C status? Approximately two-thirds will present with hand swelling. Sclerodactyly is often seen as well. There are rare reports of patients with MCTD presenting with photodistributed papulosquamous eruptions consistent with subacute cutaneous lupus erythematosus, cutaneous small vessel vasculitis, and livedoid vasculopathy. Expected results of diagnostic studies There are no characteristic histopathologic findings for MCTD. As mentioned previously, the Alarcon-Segovia criteria can be used as a fairly accurate diagnostic tool. The criteria require high titer U1-RNP antibodies defined as greater than 1: Presence of anti-Sm, anti-dsDNA, anti-Scl, and anti-centromere antibodies should steer one to reconsider the diagnosis of MCTD, as should renal or central nervous system involvement other than trigeminal neuralgia. Who is at Risk for Developing this Disease? Women are at nine times the risk of the disease as men. In contrast to SLE, there appears to be an equal prevalence between whites and blacks. What is the Cause of the Disease? It is postulated that these autoantibodies play a role in disease etiology, but this has yet to be confirmed. These autoantibodies are thought to form by a breakdown of self-tolerance during the clearance of apoptotic cells. These apoptotic cells form surface blebs filled with intranuclear material that is ultimately recognized as foreign. Others have postulated that these autoantibodies are formed via molecular mimicry of common viruses, such as the Epstein-Barr Virus EBV. Anti-U1-RNP antibodies are thought to cause vascular damage via upregulation of adhesion molecules targeting cytotoxic cells, or activation of the complement cascade Patients with MCTD also produce anti-endothelial cell antibodies and antiphospholipid antibodies that may further contribute to vascular damage. If the high resolution CT scan does not show evidence of lung fibrosis, a right heart catheterization for more careful evaluation for PAH may be warranted. Regular monitoring for cytopenias via complete blood counts, myositis via creatinine kinase and aldolase, and proteinuria via urinalysis is also recommended.

## Chapter 2 : Autoimmune Skin Diseases Programs - Brigham and Woman's Hospital

*Connective Tissue Diseases of the Skin (Basic and Clinical Dermatology) 1st Edition. by Charles M. Lapiere (Editor), T. Krieg (Editor) Be the first to review this.*

The earliest manifestations of the syndrome are: Graying at the temples Skin manifestations: The lower legs and feet, forearms and hands are most severely involved and to less extent the face and neck. Intelligence is usually normal. This type is characterized by relatively low blood glucose levels and peripheral resistance to insulin. Cataracts develop between the ages of 20 and 35 in most cases and are usually posterior and subcapsular. Other ocular defects may occur. Carcinoma has developed in a chronic leg ulcer. Death usually occurs in fourth to sixth decade, due to myocardial infarction or malignancy. Diagnosis The disease has characteristic clinical features with multi-systems involvement. The radiological changes are often striking. There may be calcification of arteries, ligaments, tendons and subcutaneous tissue with osteoporosis of the extremities, especially the legs. PROGERIA Hutchinson-Gilford syndrome This is an autosomal recessive syndrome where fibroblast survival time is decreased with increased production of hyaluronic acid that appears in urine. Clinical Manifestations Affected children usually appear normal at birth. In the first year the infant manifests with retarded growth. In the second year there is growth failure with reduced subcutaneous fat on the face and limbs. Prominent eyes and scalps veins, bird facial appearance, peaked nose and centrofacial cyanosis. Micrognathia and thin lips, large cranium with patent fontanels and frontal bossing. Skin manifestations Thin, taut and shiny skin in some areas but lax and finely wrinkled in others. Thin hair and alopecia may develop. Hypohidrosis due to decreased eccrine sweat glands. The veins are prominent and there may be easy bruising. Progeria After several years the manifestations are: Koilonychia and onychogryphosis may occur. The dentition is abnormal and delayed. There may be skeletal abnormalities, such as dystrophic clavicles and coxa vulga and joint contractures. Death usually occurs in the second decade as a result of severe generalized atheroma. Acrogeria is characterized by cutaneous atrophy and loss of subcutaneous fat particularly over the distal extremities, leading to premature aging of the extremities. Acrogeria begins at birth, where the general health and life expectancy are normal. Clinical Manifestations Short stature and low birth weight. The skin becomes dry, thin, transparent and wrinkled, especially over the hands. Poikiloderma and telangiectasia and easy bruising due to prominent veins as a result of lack of the supporting subcutaneous fat. The hands and feet may be very small. Micrognathism may be present. Premature senility due to lack of subcutaneous fat. Nails may be atrophic or thickened. The manifestation of this rare familial syndrome appears at birth. The cause is unknown, where the dermal collagen and elastin appear normal on light microscopy. Clinical Manifestations Dry wrinkled skin of the hands, feet and ventral surfaces of the trunk. General manifestations The veins are unduly prominent. There may be also mental retardation, ocular defects and poor muscle tone. Skin of diabetics may show different changes mainly: Retinal and renal disease due to microvascular damage. The histology of the skin changes resembles systemic sclerosis. The difference is a large collagen fibers, thickening of the capillary basement membrane and increased mucin. The manifestations of this syndrome are due to defect in collagen, elastic tissue or defect in the epidermal keratinocytes. The extruded materials may show inflammatory cells, red cells, microorganisms and extracellular substances, such as mucin or altered connective tissue components. Primary perforating dermatoses These include the following types: The condition is secondary to an underlying disease, such as granuloma annulare or pseudoxanthoma elasticum. Perforating dermatoses secondary to exogenous factors include: Chemicals applied to the skin topically or by intradermal injection of medications such as steroids may lead to perforating disease. Reactive Perforating Collagenosis The condition usually starts in early childhood. Clinical Features The primary lesion is skin colored small papules, which increase in size within one month to about half centimeter and then become umbulicated with keratinous plug. The lesions regress within two months leaving slight scarring or hypopigmentation. The lesions may recur again, can be elicited by trauma as a linear lesion,

or can be produced in response to cold and regress by warming the area. Treatment Topical retinoids may reduce the number of lesions. Oral isotretinoin, methotrexate, emollient creams, topical steroids under occlusion may help some cases. Perforating serpengious elastosis The age of onset ranges from 6 to 20 years. Clinical Manifestations Small horny or umbulicated papules appear mainly on the back, sides of the neck, cheek and arms. Skin lesions may be unilateral or bilateral and symmetrical. The lesions are characteristically arranged in lines, circles or segments of circles in a serpiginous pattern. The individual papules may remain small or may enlarge slightly to assume a crateriform appearance with an elevated edge and a central plug, which may leave an area of atrophic skin surrounded by smaller papules, each with a horny plug. The lesions may persist for several years but eventually involute spontaneously to leave reticulate atrophic scars, which is liable for keloid transformation. COLLOID MILIUM Colloid milium or colloid degeneration of the skin is a degenerative skin changes, characterized clinically by the development of yellowish, translucent papules or plaques on light-exposed skin and histologically by the presence of colloid in the dermal papillae. In young children, the lesions are often confined to the face, around the orbits, the backs of the hands, the back and sides of the neck and the ears, with diffuse infiltration surmounted by innumerable small papules, which may appear vesicular. Clinical features The primary lesions are small dermal papules mm in diameter, yellowish-brown and sometimes translucent, develop slowly and more or less symmetrically in irregular groups in areas exposed to sunlight. They feel soft and may release their gelatinous contents when punctured. Milia In older patients the papules are often fewer, larger and their potential distribution is much wider, although often only one or two sites are involved in each individual. Treatment CO2 laser ablation of the extensive lesions may give better cosmetic results. Superficial resurfacing by Co2 laser using topical anaesthetics as Emla creams Dermabrasion, diathermy and cryotherapy, can also give good results. Childhood systemic lupus erythematosus. Arch Dermatol ;  Perspectives in pediatric systemic lupus erythematosus. J Rheumatol ;  Systemic lupus erythematosus in a premature infant. Arthritis Rheum ;  Systemic lupus erythematosus presenting as a bullous eruption in a child. Clin Exp Rheumatol ; 6: Bullous systemic lupus erythematosus. J Am Acad Dermatol ; 7: Chronic bullous disease of childhood, childhood cicatricial pemphigoid and linear IgA disease of adults. J Am Acad Dermatol ;  Bullous dermatosis and systemic lupus erythematosus in a year-old boy. Bullous eruption of systemic lupus erythematosus. A comparative study of benign chronic bullous dermatosis of childhood and linear IgA dermatosis of adults. Br J Dermatol ; Suppl. A study of benign chronic bullous dermatosis of childhood and comparison with dermatitis herpetiformis and bullous pemphigoid occurring in childhood. Clin Exp Dermatol ; 5: Effect of pregnancy in patients with SLE. Am J Kidney Dis ; 2 Suppl. Jackson R, Gulliver M. Neonatal lupus erythematosus progressing into systemic lupus erythematosus. Br J Dermatol ;  Neonatal lupus risk to newborns of mothers with systemic lupus erythematosus. The neonatal lupus erythematosus syndrome. Mucosal involvement in systemic and chronic cutaneous lupus erythematosus. Potasman I, Bass HM. Multiple tendon rupture in systemic lupus erythematosus: Ann Rheum Dis ;

## Chapter 3 : Clinical Dermatology, 5th Edition | Dermatology | Medicine, Nursing & Dentistry | Subjects | Wil

*Mixed connective tissue disease (MCTD) is a systemic autoimmune inflammatory disorder characterized by high titer U1-RNP antibodies, and clinical and serological overlap of systemic lupus erythematosus (SLE), systemic sclerosis (SScl), and polymyositis.*

Histopathology Histopathological changes include five cardinal signs Hyperkeratosis with keratotic plugs. Atrophy of the stratum malpighii. Liquification degeneration of the basal cells. Basophilic degeneration of the collagen. Patchy perivascular lymphocytic infiltrate. Leucopenia, thrombocytopenia and biological false positive reaction BFB for syphilis may be positive. DLE may become extensive covering wide areas of the skin simulating systemic lupus but without systemic involvement of internal organs and usually shows negative LE test. The lesions are more superficial, scarring is minimal if it happens, have a shorter course than DLE, and the eruption is polymorphous. The skin lesion is sharply demarcated, greasy, scaly patches and heal without scarring or atrophy. Dry silvery scaly patches showing neither atrophy nor scarring. Treatment Preventive measures are very important. Protecting the patient from direct exposure to sunlight and using sunscreens especially on the seashores. Extreme heat and cold exacerbate the preexisting lesions. Topical or oral steroids or chloroquine helps some. The disease may affect any age group especially adult females. Immunoglobulins, predominantly IgG, but less frequently IgM and IgA, together with complement C1, C3 can be demonstrated at the dermo-epidermal junction by immunofluorescence techniques. Clinical Features General manifestations SLE is usually accompanied by vague and fleeting general symptoms such as intermittent low-grade fever, abdominal and thoracic pain, weakness, fatigue, and arthralgia. These symptoms may continue for a long time before skin lesions appear or will not be accompanied by any skin manifestations. Skin manifestations The eruption usually begins on the more exposed parts of the skin presenting with erythematous, purpuric macules and telengectasia. Skin lesions appear in about half of the cases in the form of superficial, dusky lesions on the sun exposed areas mainly on the face that give the butter fly appearance. The lesions may be unilateral but usually bilateral and symmetrical. Usually there is no atrophy of lesions as in DLE. Purpura is a common manifestation of SLE. Systemic lupus erythematosus Angioneurotic edema Fig. Systemic lupus erythematosus Systemic manifestations Fever is usually of the remitting type. Vague abdominal and chest pain, which is related to internal organs involvement. Leukopenia, anemia, and thrombocytopenia due to bone marrow depression. Polyarthritis and purpura are important signs of SLE. Systemic lupus erythematosus Dusky, erythematosus and purpuric macules Kidney involvement: Neonatal SLE is a rare syndrome occurring in infants of mothers with connective-tissue disease. Clinical Features of Neonatal SLE Skin manifestations The infants have large annular erythematous lesions or discoid lesions that may be present at birth but always develop within the first 2 months, sometimes being precipitated by sunlight. Occasionally, the lesions may be purpuric on both the face and trunk. The rash improves after months and completely clears within a year without scarring. Neonatal LE can occur in successive pregnancies. It has long been known that the LE-cell factor and antinuclear antibodies could cross the placenta and may be demonstrated in the newborn for as long as 3 months without harming the child. Mothers of babies with congenital heart disease have a one in three chance of developing SLE or other connective-tissue disease. Anti-nuclear factors can be detected in most cases and can be used for prenatal screening. Systemic manifestations Cardiac manifestations as heart block, hepatospleenomegaly, anemia, thrombocytopenia and arthritis, may be associated with the skin lesion. Renal and central nervous system involvement is rare in children but may occur. The clinical picture, course and treatment are similar to the disorder in adults. The earliest age of onset reported is 3 months. Bullous SLE may resemble chronic bullous disease of childhood. Different organs are involved mainly the kidneys showing diffuse proliferatve nephritis, central nervous system manifestations, hepatospleenomegaly and lymph nodes enlargement are common in children, affected by SLE. Albuminuria, hematuria and casts. Leucopenia, thrombocytopenia and hemolytic anemia. Biological

false positive BFP for syphilis may be positive. LE cell is a peculiar cell which is a neutrophilic polymorph nuclear leucocyte engulfing in its cytoplasm homogenized purple staining inclusion body. LE Cell may appear as rosette shape. In acute cases and during severe exacerbation: Undue exposure to sun: Symptomatic treatment Anemia must be corrected. Estimation of C-reactive protein may be helpful: Hypertension must be treated and it is generally agreed that hydrallazine can be used safely in most patients with SLE. Nephrotic syndrome or cardiac failures need diuretics. Low C3 often indicates severe renal disease. It should be noted that not all patients require steroids. Some fulminating cases have been treated with massive doses of steroids but the advantages of such therapy rarely outweigh the risks. Prednisone, 60 mg per day, is the steroid of choice initially. Once the condition appears to be under control, the dose may be reduced gradually, until maintenance dose of about mg per day is reached. It has been claimed that a single dose daily or on alternate days, before a meal or with milk before bedtime, produces fewer side effects and does not impair the therapeutic response. Prednisone and azathioprine, or prednisone and Cyclophosphamide can give also good results in renal involvement. The erythrocyte sedimentation rate ESR is no guide to the adequacy of therapy. The titers of antinuclear antibodies often persist unchanged despite clinical remission. Anti-DNA antibody and serum complement levels may be helpful in predicting exacerbation. Steroid myopathy can occur with high doses. Prolonged high dosage of corticosteroids, for example, 60 mg of prednisone daily for 6 months, is said to improve the renal lesions more than small suppressive doses. This improvement of survival is not seen in patients with raised blood urea before the onset of therapy. There is no evidence that steroids are prophylactic and that prolonged therapy will prevent the development of new features. Serum enzymes are usually normal but urinary creatine is increased Anti-convulsants for epilepsy. Chlorpromazine is a good sedative, for cases accompanied by psychosis. There is an increased risk of aspirin hepatotoxicity in SLE. Corticosteroids are required in the acute cases and should be given in adequate dosage. In mild cases the administration of chloroquine or hydroxychloroquine may allow the dose of steroids to be reduced. Antimalarials are less useful than in DLE and for long-term therapy may be dangerous causing photosensitivity. Pregnancy is not contra-indicated, as healthy live babies have been delivered from women on antimalarial therapy throughout pregnancy. Treatment of cases not responding to prednisolone Adult cases not responding to systemic steroids can be treated by the following medications: Immunosuppressive drugs have been used for adult patients not responding to corticosteroids but their use should be given to selected cases. Immuno-suppressive drugs are not given to the young age groups having SLE. It has been concluded that azathioprine added nothing to high-dose prednisone treatment in mild or moderate renal disease. Sudden withdrawal may be followed by relapse. Cyclophosphamide may be a more effective immuno-suppressant but it is more toxic than azathioprine. Tripple therapy with prednisone plus azathioprine and Cyclophosphamide had no therapeutic advantage over prednisone and azathioprine. Chlorambucil has been found helpful. Some reports claim excellent results of prednisone combined with Chlorambucil. The long-term risk of malignancy must be considered, whenever immunosuppressive drugs are used. Treatment of cases not controlled by prednisolone and immuno-suppressive drugs. Adult cases of systemic lupus erythematous that are not controlled by prednisone and immuno-suppressive drugs, can be put under the following type of treatment: Pulse therapy with 1g methylprednisone is given intravenously in ml normal saline over 4 h on three successive days to inpatients may be helpful. Vesiculobullous skin eruptions of systemic lupus erythematosus are uncommon. The clinical manifestations are similar to that of bullous pemphigoid and dermatitis herpetiformis. Histologically, the bullae are subepidermal with a neutrophilic infiltrate occasionally resulting in micro-abscesses resembling dermatitis herpetiformis. Of particular interest to the dermatologist are those reports concerning lupus erythematosus, pemphigus vulgaris, herpes gestationis and aphthosis. Since IgA, IgM and IgE antibodies do not cross the placenta in significant amounts; this phenomenon is restricted to diseases caused by autoantibodies of IgG class. Complement does not pass across the placenta. Complement can be detected in the fetus from about the 11th week of gestation. Maternal IgG is catabolized more or less completely within the first months of life, and antibody-mediated transplacental diseases can be expected to

remit spontaneously within this period. Dermatomyositis is an autoimmune connective tissue disease involving skin and muscles. Children and older age groups may be affected with the disease that may be fatal in some cases. Clinical Features Different skin and muscle manifestations appear in dermatomyositis, which may have vague signs and symptoms making diagnosis more difficult.

## Chapter 4 : Dermatology | Karolyn A. Wanat, MD

*The Dermatology-Rheumatology Connective Tissue Disease Program is a collaboration between Massachusetts General Hospital Departments of Dermatology and Rheumatology. Autoimmune diseases such as lupus, scleroderma or dermatomyositis significantly impact both the skin and musculoskeletal system.*

## Chapter 5 : Principles of Pediatric Dermatology - Chapter 37 : CONNECTIVE TISSUE DISEASES

*Autoimmune connective tissue diseases are a group of disorders in which the immune system mistakenly attacks protein-rich tissue that supports organs and other parts of the body, such as fat, bone and cartilage.*

## Chapter 6 : Dermatology | Gretchen M. Roth, MD

*Connective tissue disorders are characterised by pathology in one or more organ systems, the existence of autoimmunity in the form of autoantibody production or disordered cell-mediated immunity, vascular abnormalities such as Raynaud's phenomenon, occlusive vascular disease and vasculitis (although the pathology in not entirely the result of vascular inflammation), arthritis or arthralgia.*

## Chapter 7 : CONNECTIVE TISSUE DISORDERS | Primary Care Dermatology Society | UK

*Andrews' Diseases of the Skin: Clinical Dermatology. 12th edition, by William D. James, Timothy G. Berger, and Dirk M. Elston. Effectively diagnose and treat a wide range of skin conditions with the latest edition of the highly regarded Andrews' Diseases of the Skin: Clinical Dermatology.*

## Chapter 8 : Mixed Connective Tissue Disease (Sharpâ€™s disease)

*Perforating dermatoses include different types of skin diseases in which some tissue protrudes from the dermis. The manifestations of this syndrome are due to defect in collagen, elastic tissue or defect in the epidermal keratinocytes.*

## Chapter 9 : Lipodermatosclerosis - Wikipedia

*Usually, patients with mixed connective-tissue disease (MCTD) present with Raynaud phenomenon, which frequently represents the initial manifestation of the disease. [13] The decreasing frequency of attacks limits progressive vascular damage. The course of the entity is often mild, but the clinical.*