

Heffner VA, Lyon VB, Brousseau DC, Holland KE, Yen K. Store-and-forward teledermatology versus in-person visits: A comparison in pediatric teledermatology clinic. *J Am Acad Dermatol* 2009; 60:956-61.

Telemedicine has become an invaluable healthcare delivery tool in an ever-expanding, medically complex society. In store-and-forward (SF) teledermatology, the data and images from a patient are collected and sent to a specialist to be viewed at a later time. The role of teledermatology in the diagnosis of pediatric skin conditions has not been studied exclusively. The objective of the study was to determine the ability of a Board certified pediatric dermatologist to correctly diagnose rashes by history and digital images alone compared with direct visualization of the patient. During the 13-month period of July 2006 through August 2007, 135 patients were enrolled. Only new patients with a chief presenting complaint of either a rash or rash descriptors (eg, bumps, spots, patches) were enrolled. In the first part of the study, to evaluate the primary outcome of intrarater agreement between in-person visits and SF images, diagnosis made by the primary pediatric dermatologist by SF on history and clinical images sent by an American Board-certified pediatrician was compared with the in-person diagnosis made by the same dermatologist. In the second part of the study, to evaluate interrater agreement of SF images, a second pediatric dermatologist was asked to view the photographs and the history data sheet from each study patient at a later date and the diagnosis was compared not only to the primary dermatologist's photographic diagnosis, but also to his in-person diagnosis. Each dermatologist gave a single diagnosis. Concordance between the in-person and photographic diagnosis by the primary dermatologist was 82%, with kappa value 0.80; higher the kappa value, greater the concordance. Clinically relevant disagreement, defined as disagreement where a change in management or therapy was required, occurred in 12% cases only. Concordance between the two dermatologists receiving photographs and history alone was 73%, with a kappa photographic

interrater agreement value of 0.69. Clinically relevant disagreement occurred in 14% cases. Concordance between the two dermatologists, one viewing the patient in person and the other viewing photographs alone was 69% with clinically relevant disagreement occurring in 16% cases. Demographic factors had no statistical significance in diagnostic agreement. There were 24 cases of disagreement that occurred in the intrarater portion of the study. In six cases (25%), it was thought that a whole-body photograph instead of only close-up images would have made the diagnosis easier. In one case, poor photographic quality was identified as the cause of the missed diagnosis. On reviewing all cases with missed diagnosis, the only wrong diagnosis was on three cases of scabies misdiagnosed as atopic dermatitis.

Comment: The use of communications technology to facilitate the provision of healthcare for persons with skin diseases is an area of increasing interest and activity. Currently, the two approaches to teledermatologic communication are store-and-forward and live interactive technology (real time). Store-and-forward teledermatology uses an asynchronous approach independent of time and place, and allows for static images and clinical information to be transmitted either from physician to physician or patient to physician via the Internet or satellite. Live interactive teledermatology operates in real time via a videoconferencing link and is structured like a traditional in-office visit, allowing for skin examination with the use of special high-definition video cameras. Often live-interactive teledermatology visits integrate the use of static images for tracking of skin lesions, which demonstrates how these technologies can be used in conjunction with one another. There have been various studies in teledermatology, although pediatric patients had not been studied exclusively in this discipline.

In this study, authors evaluate the feasibility of SF teledermatology in pediatric patients. The results of this study suggest that SF teledermatology leads

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to the accurate assessment of pediatric rashes by a pediatric dermatologist. Pairing pictures from a simple point-and-shoot camera with a brief history, the dermatologists were able to correctly treat more than 80% of rashes referred to the dermatology clinic. In previous studies of teledermatology, dermatologist intrarater agreement has been in the range of 31-88%. This study showed an intrarater agreement rate of 82%, which falls into the higher end of the range presented in the adult literature.

In this study, the most common diagnosis were eczema, atopic dermatitis, molluscum contagiosum, seborrheic dermatitis, warts, insect bite, contact dermatitis etc. The concordance rate may be influenced by various factors like the disease, type of photograph, history provided, and there is concern for bias by the primary dermatologist: seeing patient photographs and later the patient in-person, one would tend to agree with one self. The primary dermatologist in this study was given a brief verbal patient history by the primary investigator, which is not strictly SF method.

The outcome of this study has practical applications within the medical community. In a large geographic area with a small number of specialists, telemedicine has proven to be a useful tool. Knowing that pediatric skin conditions can be diagnosed accurately by SF teledermatology widens treatment options. Further studies with larger sample size are warranted in this field before its widespread acceptability.

Yost JM, Do TT, Kovalszki K, Su L, Anderson TF, Gudjonsson JE. Two cases of syringotropic cutaneous T-cell lymphoma and review of the literature. J Am Acad Dermatol 2009; 61:133-8.

Syringotropic cutaneous T-cell lymphoma (CTCL) is a rare form of CTCL characterized histologically by infiltrates of atypical lymphocytes located primarily in and around hyperplastic eccrine glands and ducts. Syringotropic CTCL is categorized by the WHO-EORTC as a subtype of folliculotropic MF. The authors reviewed the literature and described two additional cases of syringotropic CTCL, presenting as erythematous to brown scaly plaques affecting trunk, limbs, face and scalp, with areas of alopecia. Biopsy showed atypical lymphocytes with syringotropism and minimal epidermotropism. Case 1 had disseminated disease in the form of lymph node involvement and was treated with the histone deacetylase inhibitor vorinostat in addition to topical bexarotene gel,

narrowband ultraviolet (UV)-B phototherapy and topical clobetasol. In addition, the patient received radiation treatment for her scalp involvement. Case 2 was treated with radiation therapy, modified Goeckerman therapy and PUVA. Authors concluded that although there is some histologic overlap between these two variants with some cases of folliculotropic CTCL with involvement of the eccrine glands and vice versa, cumulative data regarding differences in epidemiology, pathogenesis and prognosis suggest that syringotropic CTCL may merit classification as a distinct entity separate from folliculotropic MF. Further, given the dramatic differences in response to treatment between these two diseases, a more precise classification system will provide patients with more accurate prognostic information and better guide therapeutic management.

Comment: Cutaneous T-cell lymphoma (CTCL) is a malignancy of skin-homing T-cells and can have great variability in its clinical presentation. The most common variant, mycosis fungoides (MF), accounts for almost 50% of all primary cutaneous lymphomas and is characterized by proliferation of small to medium T lymphocytes with cerebriform nuclei. Folliculotropic MF is a variant of MF characterized by infiltration of atypical lymphocytes into hair follicles, often with sparing of the epidermis. Most cases show mucinous degeneration of the hair follicles, hence the term “follicular mucinosis”. In some instances, prominent infiltration of both follicular epithelium and eccrine glands occurs and is then designated as syringotropic MF according to the most recent World Health Organization (WHO) - European Organization for Research and Treatment of Cancer (EORTC) classification.

A very similar pathologic condition, termed “syringolymphoid hyperplasia” or “syringolymphoid hyperplasia with alopecia” (SLHA), occurs when there is alopecia in addition to infiltration of atypical lymphoid cells within and around the eccrine glands. There is also glandular and ductal epithelial hyperplasia, which ultimately results in luminal obliteration. It is now thought that virtually all cases of SLHA are syringotropic forms of MF.

Syringotropic MF is an exceedingly rare form of CTCL and to date only 25 cases have been reported. They can present with erythematous lesions in a patch, plaque, or papular form. Alopecia, pruritus, hyperesthesia, and anhidrosis of affected areas of

the skin have been reported, but lesions may also be entirely asymptomatic.

Histologically, the classic features of syringotropic CTCL are hyperplastic eccrine glands and eccrine ducts surrounded by a dense, syringotropic infiltrate of atypical lymphocytes with characteristic cerebriform nuclei.

Folliculotropic MF is known to have a particularly poor prognosis when compared with other MF subtypes. In comparison, all reported cases of syringotropic CTCL have had a benign, chronic course, limited exclusively to cutaneous involvement. One review of 20 cases of syringotropic CTCL indicated that the disease had a prognosis similar to chronic forms of classic MF, with 10-year survival ranging from 83 to 97% depending on the extent of cutaneous involvement. There is only a modest difference in sex and the age of onset between these two variants, with male to female ratio of 5:2 in syringotropic MF versus 3:1 in folliculotropic MF. As syringotropic CTCL are reported, it is becoming clear that it does not have the same predilection for the head and neck that typifies folliculotropic MF. Including these two new cases, more than 50% of patients with syringotropic disease described in the literature had lesions on the trunk, hips, breasts, buttocks or groin, suggesting that clinical presentation of syringotropic CTCL closely parallels that of classic MF.

Although a first-line treatment regimen for syringotropic CTCL has yet to be established, many of the standard topical therapies for classic MF have previously been proven ineffective because of poor levels of penetration to the reticular dermal infiltrates. Consequently, most case studies report local radiotherapy to be most effective in treating local disease. Both phototherapy and photochemotherapy have also been widely tried in treating syringotropic disease, however, with mixed results. Aggressive treatment is typically not required because of the indolent course of the disease.

Poitrine FC, Revuz JE, Wolkenstein P, Viallette C, Gabison G, Pouget F, Poli F, Faye O, Garin BS. Clinical characteristics of a series of 302 French patients with hidradenitis suppurativa, with an analysis of factors associated with disease severity. J Am Acad Dermatol 2009; 61:51-7.

Hidradenitis suppurativa (HS), defined as chronic suppuration of inverse areas, is characterized

by recurrent, painful, deep-seated nodules and abscesses of apocrine gland-bearing skin. Factors associated with severity of HS are not known. Until now, potential clinical differences between the sexes and associated with factors such as obesity, associated follicular disease and atypical locations were unknown. Gender, smoking, obesity and a family history of HS are presumed to be linked to severity. During the study period from 1998-2006, 302 patients were included, 232 female and 70 male and disease severity was assessed by the Sartorius severity score. Maximal pain score and maximal suppuration score were assessed by the patient for the preceding month, with a numeric scale from 0 to 10. The number of painful days per month and the number of days with suppuration were evaluated by the patient in four categories: none, less than 15 days, greater than or equal to 15 days, or constant. Characteristics of the patients and their diseases were described, and compared between men and women. The median age was 30.4 years at the time of evaluation and 20.0 years at the beginning of disease. Atypical locations (ears, chest and other atypical locations [eg, abdomen, legs]) were more common in men than in women (47.1% vs 14.8%; $P < 0.001$). The front part of the body (i.e. groin and breasts) was predominantly involved in female patients, whereas involvement of the back of the body (i.e. buttocks and perianal area) was a hallmark of male patients. Men also had more severe disease (median Sartorius score: 20.5 vs 16.5; $P = 0.02$). Increased body mass index ($P < 0.001$), atypical locations ($P = 0.002$), a personal history of severe acne ($P = 0.04$), and absence of a family history of HS ($P = 0.06$) were associated with an increased Sartorius score. The Sartorius score was highly correlated with the intensity and duration of pain and suppuration (all P values < 0.001).

The author concludes that these data indicate significant association between the severity of HS and several clinical and behavioral factors. Prospective studies are needed to confirm the prognostic role of these factors. The recognition of a wide spectrum of severity has an impact on the treatment of patients with HS; an exclusively surgical approach does not seem justified.

Comment: Hidradenitis suppurativa was first described as a distinct entity in 1839. It is a disorder of the terminal follicular epithelium in the apocrine gland-bearing skin. Hidradenitis suppurativa is

characterized by comedo like follicular occlusion, chronic relapsing inflammation, mucopurulent discharge, progressive scarring and causes impairment of the quality of life. This study provides comprehensive clinical description, objective and subjective severity parameters of HS with a good sample size of 302 patients. The author has used two scoring systems; Hurley classification and Sartorius score

Sartorius score was used to assess severity even though it had not been formally validated. This score has been used in several studies and shows a good correlation with impairment of quality of life and a good sensitivity to change while on treatment. A strongly positive association was found between the Sartorius score and the classic Hurley classification and, more importantly, with the markers of disease burden: intensity of pain and suppuration. These observations suggest that the Sartorius score may well reflect the severity of HS. Correlation of these markers of disease burden with impairment of the quality of life has already been established.

In contrast to most previous reports, there was a high proportion of "benign" involvement in this series: the most severe form (i.e. Hurley grade III) represented only four per cent of patients, whereas 70% were Hurley grade I. Increased incidence in females, association with BMI, negative association with family history found in this study have been described earlier. A relationship between obesity and the severity of HS has been considered, but never established. Men had more severe diseases as assessed by the Sartorius score, but this was linked to the presence of atypical locations and a personal history of severe acne. The presence of a pilonidal cyst, which was more frequent in men, was not significantly linked to severity, but there was a trend. Thus, a severe form of HS can be tentatively delineated. This form, which occurs more frequently in men, is characterized by the association of follicular diseases (i.e. acne and pilonidal cyst) and lesions in atypical locations.

Smoking status was not associated with the severity of HS. It is a well recognized factor associated with the risk of HS and considered by some authors to be a true risk factor. Smoking seems to be a triggering factor but, according to these results, has no role in aggravating HS. As the treatment of this chronic condition is frustrating, knowledge of these risk factors may help modify its course and improve quality of life of patients.

Adler RS, D'Aquilante DA, Kovarik CL. Antiretroviral-induced genital ulceration. J Am Acad Dermatol 2009; 61:164-165.

A 45-year-old HIV positive male on highly active antiretroviral therapy (HAART), with a viral load of 300, CD4 count of 550 presented with penile ulcers since 1999. On examination, there were several slightly erythematous ulcers, up to 2.5 cm in diameter, on the penile shaft and glans. Inguinal lymphadenopathy was appreciated, but no other oral or cutaneous lesions were present. Biopsies from the ulcers showed epidermal ulceration with associated spongiosis and a dense mixed dermal infiltrate. Work up for sexually transmitted infection and autoimmune cause was negative. The patient gave a history of ulcers which completely resolved when he stopped his antiretroviral medications. Drug-induced ulceration was suspected and his antiretroviral medications were discontinued in July 2007, with complete healing of the ulcers by August 2007. The patient had been on two HAART regimens in the past (comprising of tenofovir, abacavir, lamivudine, atazanavir, and ritonavir), both of which resulted in ulcers. Medications in common to both regimens were ritonavir and lamivudine. The authors concluded that antiretroviral medications should be considered as a possible cause of genital ulceration in HIV patients on HAART after a complete infectious, neoplastic, and inflammatory/rheumatologic workup has been done.

Comment: Since the introduction of HAART, cutaneous drug reactions have been reported with all antiretroviral medications; however, only zalcitabine, nevirapine, and saquinavir have been described as causing mucocutaneous ulcerations. These ulcerations may occur even months after initiation of therapy, typically resolve after cessation, and recur on rechallenge.

Neither of the medications used in this patient have been previously described as causing persistent genital ulcerations. Elucidating the time course of therapy is critical in the diagnosis of medication-induced genital ulceration.

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