confounders include increased exposure to tobacco smoke and increased respiratory symptoms before the study in the children randomly assigned to the probiotic group. Additional studies are required to assess probiotic use in children with a personal or family history of atopic disease.

CONCLUSIONS. The traditional Chinese herbal medication XFS might be an alternative choice of therapy for severe, refractory, extensive, nonexudative atopic dermatitis.

REVIEWER COMMENTS. Severe and widespread atopic dermatitis can be frustrating to treat for patients, parents, and physicians. Patients often ask their physicians if there are alternative approaches to controlling atopic diseases. XFS is a common Chinese herbal preparation of 12 herbs, some with known anti-inflammatory effects, that might provide a complementary option for adults and children who require systemic steroids to control their eczema flares. However, more scientific evaluation of XFS to determine its mechanism of action, safety profile, applicability, and palatability need to be considered before widespread use is accepted.

URL: www.pediatrics.org/cgi/doi/10.1542/peds.2011–2107AAAA

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The Efficacy and Safety of a Chinese Herbal Product (Xiao-Feng-San) for the Treatment of Refractory Atopic Dermatitis: A Randomized, Double-Blind, Placebo-Controlled Trial
Cheng HM, Chiang LC, Jan YM, Chen GW, Li TC.
Int Arch Allergy Immunol. 2011;155(2):141–148

PURPOSE OF THE STUDY. To determine if the Chinese herbal product Xiao-Feng-San (XFS) taken orally could significantly improve symptoms of severe intractable atopic dermatitis.

STUDY POPULATION. A total of 71 Taiwanese subjects (age range: 8.4–22.6 years [median: 13.1 years]) with history of severe, refractory atopic dermatitis and poor response to eczema medications (topical steroids, oral antihistamines) were enrolled.

METHODS. This was a prospective, double-blind, placebo-controlled trial in which patients were randomly assigned at a ratio of 2:1 to receive XFS or placebo over 8 weeks. There were 47 (median age: 12.2 years) given XFS and 24 (median age: 13.6 years) given placebo. Participants were matched according to gender, height, weight, BMI, age, duration of illness, and symptom scores. Patients were given varying doses depending on age. Laboratory studies were performed and total lesion score, erythema score, surface damage score, pruritis score, and sleep scores were calculated at 4-week intervals up to 12 weeks.

RESULTS. A total of 56 subjects completed the entire study. There was a statistically significant improvement in total lesion scores among those in the treatment group compared to those of the placebo group (79.7 ± 5.8% vs 13.5 ± 7.64%; P < .001). There was also statistically significant improvement in all symptom scores for those on treatment compared to those on placebo. Four weeks after the treatment was discontinued, the mean improvement in the clinical lesion score for the XFS group was still significantly better than that of the placebo group. Patients reported no adverse effects except unpalatability for some. Treatment did not affect total serum immunoglobulin E level, eosinophil counts, or interleukin 5, interleukin 13, or eosinophil cationic protein levels.

URL: www.pediatrics.org/cgi/doi/10.1542/peds.2011–2107BBBB

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URGENT INSIGNIFICANT IMMUNODEFICIENCY, HIV, AND INFECTIOUS DISEASES

Chronic Mucocutaneous Candidiasis in Humans With Inborn Errors of Interleukin-17 Immunity

PURPOSE OF THE STUDY. Chronic mucocutaneous candidiasis disease (CMCD) is characterized by recurrent or persistent infections of the skin, nails, and oral and genital mucosa caused by Candida albicans and sometimes Staphylococcus. Previous studies have shown that interleukin 17 (IL-17) receptor-deficient mice were more susceptible to oropharyngeal candidiasis and staphylococcal infections of the skin. The purpose of this study was to assess if findings in the mouse model also applied to humans.

METHODS. Candidate gene sequencing was performed on a child with C albicans in the neonatal period and Staphylococcus aureus dermatitis at 5 months of age and a family from Argentina with autosomal dominant pattern of CMCD inheritance. Sequences of IL-17-related genes and receptors from affected people were compared with those of family members and controls. Additional experiments were performed by incubating fibroblasts from an affected child with recombinant IL-17A and IL-17F homodimers and heterodimers.

RESULTS. The initial child was found to be homozygous for a mutation in the IL17RA gene that was not found in any of the controls. The IL-17RA protein was not detected on the surface of fibroblasts, CD4+ T cells, CD8+ T cells, or monocytes from the patient. The patient’s fibroblasts did not respond to any of the 3 IL-17 cytokines. In the family...
studied, a heterozygous missense mutation was found in the *IL17F* gene. The mutant allele was found in 2 apparently healthy family members, which suggests incomplete clinical penetrance, and in all of the affected members of the kindred. This mutant protein was tested in a cell line and was nonfunctional.

**CONCLUSIONS.** Mutations in IL-17–family genes that cause functional deficiency of this pathway are associated with CMCD.

**REVIEWER COMMENTS.** This is an excellent example of bench-to-bedside medicine and illustrates the utility of murine models of immunity in the search for causes of human disease. These findings provide definitive evidence that IL-17A and IL-17F are essential for protective immunity to *C. albicans* and, to a lesser extent, *S. aureus* in the nails, skin, and oral and genital mucosa and provide new opportunities for designing novel treatments for this chronic immunologic disorder. It is also important to consider that elevated levels of IL-17 have been associated with various chronic inflammatory conditions, which raises the possibility that anti–IL-17 treatment strategies now under development could lead to increased susceptibility to infections with *C. albicans* or *S. aureus*.

URL: www.pediatrics.org/cgi/doi/10.1542/peds.2011–2107CCC

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**Clinical Disease Caused by Klebsiella in 2 Unrelated Patients With Interleukin 12 Receptor β1 Deficiency**


**PURPOSE OF THE STUDY.** To describe a new infectious disease phenotype in patients who inherit mutations in the interleukin 12 (IL-12) signaling pathway.

**STUDY POPULATION.** In this report the authors documented sepsis with *Klebsiella pneumoniae* in 2 unrelated patients with complete defects in the IL-12 receptor β1.

**METHODS.** This was a chart review with case reports.

**RESULTS.** The first patient was born to unrelated parents and developed BCGitis after BCG vaccination in infancy followed by non typhoidal salmonellosis. Both infections were difficult to treat despite multiple appropriate antimicrobial agents administered over 26 months. This was followed by development of disseminated *Mycobacterium bovis* infection, which also responded poorly to multidrug antimicrobial agents along with interferon γ therapy. The patient’s condition worsened, and he developed...
Chronic Mucocutaneous Candidiasis in Humans With Inborn Errors of Interleukin-17 Immunity

Douglas F. McMahon and James E. Gern

*Pediatrics* 2011;128;S140

DOI: 10.1542/peds.2011-2107

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*Pediatrics* 2011;128;S140
DOI: 10.1542/peds.2011-2107CCCC

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