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ranging from 4 to 8 weeks. Clinical features include cervical adenitis, vomiting, and diarrhea. A patient with HIDS may present to a dermatologist with a maculopapular rash, with petechiae and purpura that appear during a febrile attack. Generalized lymphadenopathy and rash are very common in these patients.

Distinctive laboratory features include an elevated IgD but this elevation is not present in all HIDS patients. The gene for HIDS has been mapped to chromosome 12 and at least 8 different mutations or deletions have been seen, but the syndrome is most likely to occur in people with Dutch or French ancestry, Dr. Edwards said.

► **Tumor Necrosis Factor–Receptor Associated Periodic Syndrome (TRAPS).** Children with TRAPS may have a lifelong history of febrile episodes that last 2-3 weeks at a time, but the febrile episodes only occur 2-3 times per year.

Conjunctivitis and raised red lesions distinguish TRAPS from other familial periodic fever syndromes. One study of 25 TRAPS patients showed that 21 (84%) had erythematous patches, including both wavy and circular lesions (N. Engl. J. Med. 2001;345:1748-57). Other clinical features of TRAPS include myalgia, arthralgia, and abdominal pain.

Skin manifestations are much more common with TRAPS than with the other familial periodic fever syndromes. "Almost all of these children will have skin lesions that may persist even when the fever is gone," Dr. Edwards noted.

When a febrile episode occurs, TNF receptors are suppressed, which creates an uncontrolled inflammatory response. Consequently, TNF inhibitors can be used to treat these patients, Dr. Edwards said.

► **Muckle-Wells Syndrome/Familial Cold Urticaria.** These two syndromes are both associated with mutations of the CIAS1 gene family. Mutations in these genes lead to autoinflammatory syndromes in which large numbers of cytokines are generated, which means that amyloidosis is very frequent in these individuals.

Patients with Muckle-Wells syndrome (MWS) generally present with urticaria and progressive sensorineural loss and deafness. Because MWS is a disease of dominant genes, the parent may show signs of hearing problems, which should prompt clinicians to include MWS in the differential diagnosis of recurrent urticaria and fever.

By contrast, patients with familial cold urticaria will present not only with urticaria and wheals, but with complaints of painful joints, chills, and fever. Febrile episodes in patients with familial cold urticaria generally occur several hours after exposure to cold. Both syndromes are associated with German, English, French, and North American ancestry. ■

New Terbinafine Formulation Knocks Out Tinea

CHICAGO — A new oral formulation of the antifungal drug terbinafine significantly improved tinea capitis in children aged 4-12 years compared with griseofulvin oral suspension, based on efficacy data from 1,286 children in the largest study of the medication to date.

These findings were presented in a poster by Dr. Sheila Friedlander at the annual meeting of the Society for Pediatric Dermatology.

Children with confirmed positive cultures for tinea capitis who were randomized to receive terbinafine had a significantly higher complete cure rate (combined mycologic and clinical cure rates) after 6 weeks of daily treatment and 10 weeks of follow-up, compared with those who received griseofulvin (45% vs. 39%), said Dr. Friedlander, a pediatric dermatologist at the University of California, San Diego Medical Center.

The new terbinafine formulation (Lamisil oral granules) consists of coated granules that can be sprinkled on food so children can swallow them easily. Both terbinafine and griseofulvin are dosed by body weight. The study was supported in part by Novartis Pharmaceuticals Corp.

Adverse event rates were similar between the two groups. About half of the patients in each group reported at least one adverse event, but almost all were mild or moderate; only 1.6% of the terbinafine patients and 1.2% of the griseofulvin patients discontinued their medications because of adverse events. The most common complaints included vomiting, diarrhea, headache, and abdominal pain.

The mycologic cure rate alone was significantly higher in the terbinafine group compared with the griseofulvin group (62% vs. 56%). The clinical cure rate alone was higher, but not significantly higher, in the terbinafine group compared with the griseofulvin group (63% vs. 59%). Terbinafine was most effective against *Trichophyton tonsurans*, which is the organism most often associated with tinea capitis, Dr. Friedlander and her associates wrote.

—Heidi Splete