protection against bacterial pathogens and compensates for B-cell dysfunction.

In summary, CVID is a heterogeneous immunodeficiency syndrome that spans a broad clinical spectrum. A small subset of patients with a presumptive diagnosis of CVID is spared recurrent infections despite hypogammaglobulinemia and impaired antibody responses to vaccines. Identification and registry of these patients will lead to broader awareness of CVID and will offer an opportunity to study potential innate mechanisms harnessed by the immune system to compensate for cognate defects.

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THE PANCAKE SYNDROME (ORAL MITE ANAPHYLAXIS) BY INGESTION AND INHALATION IN A 52-YEAR-OLD WOMAN IN THE NORTHEASTERN UNITED STATES

Oral anaphylaxis to ingested mites is a severe and potentially life-threatening reaction that has been reported primarily in semitropical and tropical climates. We describe a patient from the northeastern United States who experienced anaphylaxis on 2 separate occasions after ingesting and inhaling cooking vapors from a commercial pancake mix contaminated with the house dust mite *Dermatophagoides farinae*. The diagnosis was made by visual inspection of and skin testing with the pancake mix.

A 52-year-old female resident of Danvers, Massachusetts, experienced her first episode of eye swelling and lip tingling shortly after eating pancakes made from a buttermilk pancake mix (Krusteaz Pancake Mix; Continental Mills, Seattle, Washington) that had been stored in her kitchen cabinet for 2 years. This reaction was initially attributed to an unknown environmental allergen, because the patient had a history of allergy to dust mites, pollen, mold, and dander, but no history of food allergy. Three months later, while cooking pancakes from the same mix, she experienced wheezing, diffuse redness, and facial swelling that again required emergency department care. The patient was not aspirin sensitive, but was taking a β-blocker for hypertension.

On allergy evaluation, she had positive skin prick test results (15-mm wheal) to house dust mites (*D farinae* and *Dermatophagoides pteronyssinus*; ALK Labs, Round Rock, Texas) and a 9-mm wheal and a 25-mm flare to the pancake mix. Skin prick test results to wheat were negative, and the patient had no history of problems after eating wheat-based products, including pancakes. Although grossly normal in appearance, microscopic examination of the pancake mix revealed a thriving colony of live mites, easily seen under low power (a video is available at http://www.annallergy.org). Larry Arlian, PhD, at Wright State University, Dayton, Ohio, confirmed the species as *D farinae*, and a 2-site monoclonal antibody enzyme-linked immunosorbent assay (Indoor Biotechnologies Inc, Charlottesville, Virginia) measured a Der f1 concentration of 322 μg/g of the pancake mix.

A review article[1] in this journal described relatively few published cases of anaphylaxis from oral mite ingestion. To our knowledge, Erben et al[2] reported the first case of anaphylaxis to mite-contaminated food in 1993, when they described a 48-year-old house dust mite–allergic man who developed anaphylaxis after eating beignets contaminated with *D farinae*. Blanco et al[3] have described patients from Spain and Venezuela, some of whom were aspirin sensitive, who experienced severe allergic reactions, some repeated and life threatening, to foodstuffs that contained high quantities of mites. Because most of their patients reacted to mite-contaminated pancake flour, they labeled this condition the "pancake syndrome." Other reports[4–6] describe additional cases of anaphylaxis after the ingestion of grains contaminated with mites, often storage mites.

In April 2006, a "Dear Abby" letter[7] described a 14-year-old boy who experienced dyspnea and cyanosis 10 minutes after eating an outdated pancake mix. This letter cited a 2001 publication[8] by medical examiners from Charleston, South Carolina, that described the death of a 19-year-old asthmatic man after the ingestion of pancakes made from a packaged mix that had been opened and stored in a cabinet for approximately 2 years. Autopsy showed laryngeal edema and hyperinflated lungs with mu-

Disclosures: Dr Miller is owner and CEO of Mission:Allergy, Inc, a manufacturer and distributor of products for allergen avoidance.

This Letter has an online supplement (a video) that is accessible from this issue's Table of Contents at http://www.annallergy.org.

cous plugging consistent with an asthma-related death. The pancake mix was analyzed and found to contain 700 CFU of mold per gram of mix. The researchers presented this case as an unusual case of anaphylaxis to mold-contaminated pancake mix in a patient with mold allergy.

However, anaphylactic reactions in mold-allergic patients to molds in fermented foods, such as cheese, dried fruits, yogurt, and wine, are quite rare, if even existent. The term *farinae* (as in *D farinae*) means "wheat," and wheat products can be used as culture media for mites. High humidity favors the growth not only of mold but also of mites. In our opinion, the "Dear Abby" case and the death of the 19-year-old man in Charleston were more likely because of mites rather than mold in the pancake mix.

The message from our case report and others cited in the literature is that anaphylaxis from oral ingestion of mites—or from inhalation of mite-contaminated grain mixtures—is a severe, potentially lethal, allergic condition that may be frequently undiagnosed. Although the present patient was not aspirin sensitive, her use of a β-blocker may have increased the severity of her reaction. Most previous cases have been reported in tropical and subtropical countries. Our case from the northeastern United States suggests that this problem may be more prevalent in temperate climates than previously recognized. A female dust mite can lay 60 eggs in its 80-day lifetime. With it taking only weeks for a dust mite to reach maturity, contamination of a wheat product with only a few mites can, in conditions of adequate humidity, lead to a thriving culture within months.

Flour-based products should be stored in a refrigerator or freezer after purchase. Allergy specialists should microscopically examine suspected foodstuffs in cases of unexplained or idiopathic anaphylaxis.

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ADOLESCENT PRESENTATION OF X-LINKED LYMPHOPROLIFERATIVE DISEASE

X-linked lymphoproliferative disease (XLP; OMIM 308240) is characterized as a rare immunodeficiency that presents with Epstein-Barr virus (EBV) infection and has an average age of onset of 2.5 years.[1] The molecular characterization of XLP has led to improved and rapid diagnosis and advanced our understanding in immune responses and development. Gene sequencing in patients with XLP has revealed *SAP/SH2D1A* gene (chromosome Xq25 encodes the SAP protein) mutations in 60% of patients.[2] Recently, X-linked inhibitor-of-apoptosis deficiency has been reported to cause the clinical phenotype of XLP in 3 families.[3] Since SAP is expressed in $CD8^+$ T cells, a rapid screening test was developed recently for detection of SAP protein by flow cytometry.[4]

The patient was a previously healthy 16-year-old boy who presented with a 2.5-week history of cough, sore throat, and fever. Tonsillar exudates, lymphadenopathy, and a positive Monospot test result were documented. Five days later, he was admitted to a local hospital for dehydration and was treated with intravenous fluids and corticosteroids. After 4 days, he was transferred to a tertiary care hospital because of hepatic failure. On examination, he was febrile and appeared ill. He displayed respiratory distress, ascites, hepatosplenomegaly, jaundice, and generalized lymphadenopathy. The patient developed respiratory distress that required assisted ventilation and coagulopathy. He was treated with antibiotics, ganciclovir, and intravenous immunoglobulin. The patient had no history of recurrent infections, autoimmune disease, or prior known EBV infection. The family history was unremarkable in first-degree relatives, but a maternal great-uncle died at the age of 9 years of uncharacterized meningoencephalitis and pneumonia (Fig 1A).

EBV titers indicated an acute infection (EBV early antigen, EBV viral capsid antigen IgG, and EBV viral capsid antigen IgM positive and EBV nuclear antigen IgG negative). Polymerase chain reaction for EBV demonstrated 529,500 copies/mL. Immunoglobulin levels were polyclonally elevated, and flow cytometry from the peripheral blood demonstrated elevated B-cell counts and an inverted $CD4^+/CD8^+$ T-cell ratio of 0.7.

On the third day of hospitalization, while awaiting liver transplantation, the patient developed pulmonary hemorrhage that failed to respond to aggressive intervention. The patient's course deteriorated rapidly and a cardiopulmonary arrest ensued. Resuscitative efforts were withheld according to the family's request and the patient died.

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