the effect is smaller in magnitude than the findings of Jackson et al,[7] who more accurately measured viral etiology. Misclassification resulting from the imprecision in defining RSV and RV seasons, with RV occurring during the RSV season but RSV not occurring during the late spring and fall RV season, as well as our inability to capture RV-wheezing illnesses for which medical care was not sought in the RV-predominant group, would result in underestimation of the differential risk between the 2 seasons.

In conclusion, although bronchiolitis diagnosis during infancy was associated with an approximately 2-fold increased risk of early childhood asthma, this risk differed by season of bronchiolitis. Bronchiolitis occurring during RV-predominant months was associated with an estimated 25% increased risk of early childhood asthma compared with RSV-predominant months. This work supports recent findings that early RV-wheezing illnesses are associated with higher risk of subsequent asthma than other viruses.[7] Because of higher rates of bronchiolitis during the winter virus season, the proportion of associated asthma after winter virus bronchiolitis, however, is greater.

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Novel presentation of Omenn syndrome in association with aniridia

To the Editor:

Omenn syndrome (OS) is an autosomal recessive combined immunodeficiency characterized by infiltration of the skin and gastrointestinal tract by activated oligoclonal T lymphocytes.[1] Patients are profoundly hypogammaglobulinemic, with few or absent circulating B lymphocytes but high IgE levels. The most common causes of OS are hypomorphic mutations of the recombination-activating genes *(RAG)*.[1] Mutations of the Artemis *(DCLRE1)*, IL-7 receptor α, *RMRP*, IL-2 receptor γ, and *CHD7* genes can also result in the OS phenotype. *RAG1* and *RAG2* are located on chromosome 11p13 in close proximity to a cluster of genes that are deleted in microdeletion 11p13 syndrome. This syndrome, also known as Wilms tumor susceptibility, aniridia, genitourinary abnormalities, and mental retardation (WAGR), arises from deletions encompassing paired box gene 6 *(PAX6)* and Wilms tumor 1 gene *(WT1)*. *PAX6* encodes a transcription regulatory protein and is essential for the development of multiple tissues in the eye, including the iris, lens, and neuroretina. Heterozygous *PAX6* mutations often cause hereditary aniridia.[2] Additionally, *PAXNEB* (a *PAX6* neighbor gene) mutations on 11p13 have been studied for a possible association with aniridia.

We describe a novel presentation of OS associated with aniridia. We show evidence that this unique phenotype results from compound heterozygosity for *RAG* point mutations on the maternal allele along with a contiguous deletion encompassing both *RAG* and *PAX6* genes on the corresponding region of the paternally derived chromosome 11.

The patient was a 3-month-old boy admitted for treatment of a purulent pericardial effusion with blood cultures positive for *Achromobacter xylosoxidans*. Previously, he had been a full-term infant with aniridia who had an erythematous peeling rash on his entire body during the first week of life. This was treated as eczema, with temporary improvement followed by progressive worsening. He experienced poor weight gain through his first 3 months. He was the only child of nonconsanguineous healthy parents of Western European descent. The family history was negative both for immunodeficiency and aniridia.

The physical examination was notable for severe erythema with diffuse skin peeling, seborrheic dermatitis, and paucity of

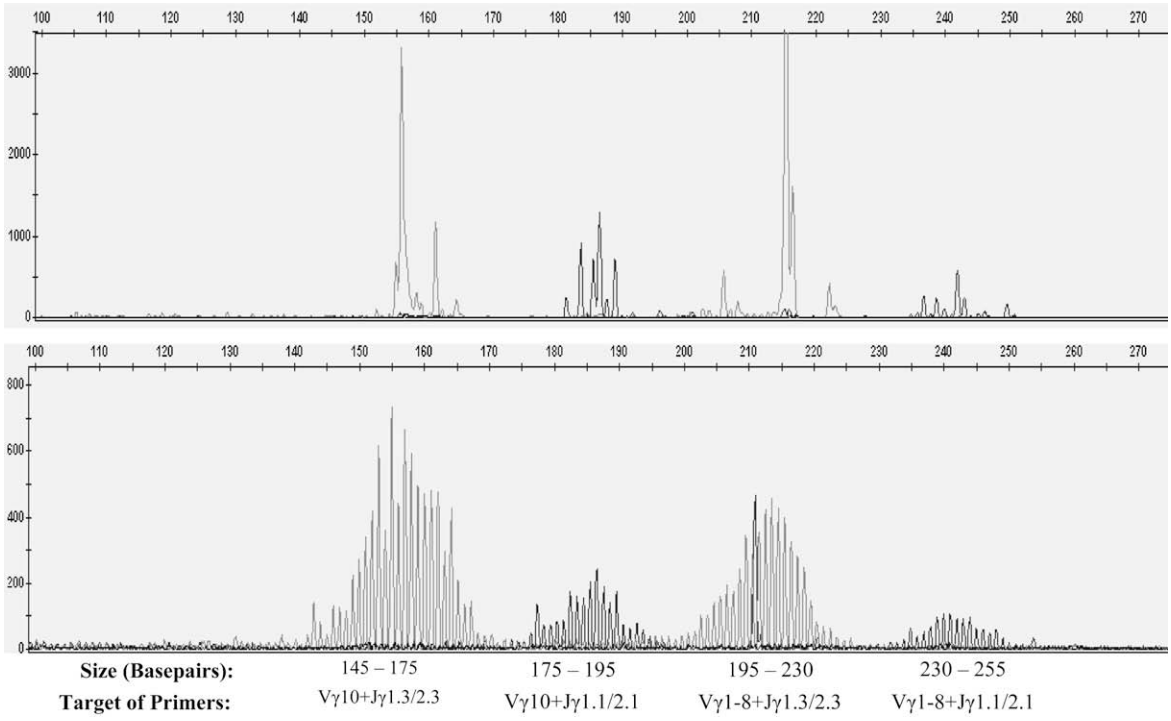

FIG 1. T-cell receptor γ gene rearrangement clonality analysis. Multiplex PCR amplification with primers to the variable and joining regions of T-cell receptor γ was used to evaluate the V-D-J rearrangements for diversity. PCR product sizes are shown on the x-axis, and peak fluorescence is shown on the y-axis. *Top row* (patient with OS): Oligoclonal pattern indicative of limited TCR γ diversity. *Bottom row* (control subjects): Normal distribution of T-cell receptor γ rearrangement indicative of adequate diversity.

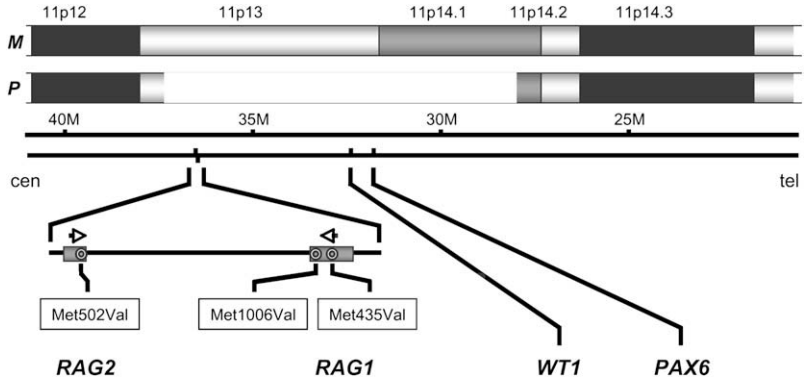

FIG 2. Schematic representation of chromosome 11p with the deletion and *RAG1*/*RAG2* gene mutations *(circles)* in the patient's paternally *(P)* and maternally *(M)* derived alleles. The deletion encompasses approximately 9.5 Mb, extending from 11p12 to 11p14.1 (chr11:27,956,661-37,509,625; hg18). The median spacing of probes on the microarray is 9 kb.

hair. Aniridia was noted. A 2/6 holosystolic murmur was present. The rest of the physical examination was normal. Of note, there were no genital abnormalities, lymphadenopathy, or organomegaly. Initial laboratory studies included a white blood cell count of 81,970 cells/μL with 15% segmented neutrophils, 73% lymphocytes, 8% eosinophils, and 2% basophils; a hemoglobin count of 9.1 g/dL; a hematocrit value of 30%; a platelet count of 263 × 10^3/μL; an IgG level of 41 mg/dL; an IgA level of 9 mg/dL; an IgM level of 8 mg/dL; and an IgE level of 196 mg/dL. Additional studies revealed an absolute lymphocyte count of 59,840 cells/μL, an absolute $CD3^+$ count of 50,226 cells/μL (96%), an absolute $CD4^+$ count of 18,737 cells/μL (32%), an absolute $CD8^+$ count of 36,142 cells/μL (62%), an absolute $CD19^+$ count of 37 cells/μL (0%), and an absolute $CD16^+/CD56^+$ count of 1,107 cells/μL (2%). The patient's lymphocytes were unresponsive to PHA, concanavalin A, and pokeweed mitogen. His karyotype was normal 46, XY. Maternal engraftment was excluded by means of fluorescent *in situ* hybridization. PCR amplification of lymphocyte Vγ–Jγ segments showed oligoclonality with profoundly limited T-cell diversity, which is consistent