Of the remaining patients, 26 children (16%) were found to have an immunologic abnormality, and a primary immunodeficiency was identified in 17 of these patients (10%). The majority of the patients with an identified immunodeficiency had a defect associated with antibody production, whereas there were also patients with complement defects (n = 3), MyD88 deficiency (n = 1), and congenital asplenia (n = 1). Importantly, patients presenting with invasive pneumococcal disease aged >2 years had a much higher risk of having a primary immunodeficiency (26% vs 2% in patients aged <2 years at presentation).

CONCLUSIONS. Patients presenting with invasive pneumococcal disease should undergo an immunologic evaluation, particularly those who present with disease who are aged >2 years.

REVIEWER COMMENTS. This study is the first prospective evaluation of immunized children presenting with invasive pneumococcal disease. The incidence during the first 4 years of the study (2005–2008) was 23 per 100,000 for children aged <2 years and 5 per 100,000 for children aged ≥2 years. Universal immunization with 7-valent pneumococcal conjugate vaccine was initiated in France in 2006, and this was switched to 13-valent pneumococcal conjugate vaccine in 2010, with vaccination coverage reaching ~93% by 2011. The majority of the serotypes found in the patients with invasive pneumococcal disease were not found in the 7-valent pneumococcal conjugate vaccine, suggesting that “herd” immunity is at play regarding the serotype exposure among susceptible children. This study may actually underestimate the frequency of invasive pneumococcal disease because it did not include children missed who may have died of this infection. The take-home message from the study is that any child who develops invasive pneumococcal disease should undergo an immunologic evaluation focused on leukocyte count and differential, immunoglobulin levels, and complement (classic and alternative pathway) activity. In addition, these patients should be assessed for congenital asplenia as well as anatomic abnormalities that could be associated with infectious susceptibility. If results of all these studies are unrevealing, referral to a clinical immunologist with expertise in primary immunodeficiency disorders should be considered for evaluation of toll-like receptor function.

Deficiency of Innate and Acquired Immunity Caused by an IKBKB Mutation


PURPOSE OF THE STUDY. The goal of this study was to characterize a form of severe combined immunodeficiency (SCID) that has previously been described.
Deficiency of Innate and Acquired Immunity Caused by an IKBKB Mutation

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