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Insights into the clinical spectrum of selective IgA deficiency: Data from two centers

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Abstract
Introduction: Selective IgA deficiency (sIgAD) is the most common primary immunodeficiency, yet its clinical presentation ranges from asymptomatic cases to individuals suffering from recurrent infections, allergic manifestations, and autoimmune disorders. Limited data exist regarding the immunological and clinical profiles of pediatric patients with sIgAD in Türkiye.
Methods: We conducted a retrospective analysis of 45 pediatric patients (20 females and 25 males) diagnosed with sIgAD and followed at two tertiary care centers. Demographic features, allergic and autoimmune comorbidities, and immunological parameters were evaluated. Lymphocyte subset analyses and immunoglobulin subclass levels were recorded. Associations between IgG3/IgG4 subclass deficiencies and infection frequency were assessed using the Mann-Whitney U test.
Results: The median current age was 102 months (range: 48-204), with a median age of symptom onset at 24 months (range: 1-186), referral at 88 months (range: 6-199), and diagnosis at 87 months (range: 48-192). A history of at least one allergic disease, including asthma, allergic rhinitis, and/or atopic dermatitis, was present in 66.7% of patients. Autoimmune conditions were identified in 13.3%, including Hashimoto's thyroiditis, vitiligo, and immune thrombocytopenic purpura. No statistically significant differences in the frequencies of upper respiratory tract infections, pneumonia, otitis, or viral infections were observed between patients with low versus normal/high IgG3 or IgG4 levels (all $P > 0.05$).
Conclusion: Our findings highlight the high prevalence of allergic diseases and the clinical heterogeneity of sIgAD in children. Moreover, isolated IgG3 or IgG4 subclass deficiencies may not independently influence infection susceptibility. Longitudinal studies are warranted to better define the prognostic role of immunoglobulin subclasses in pediatric sIgAD.

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Introduction

Selective immunoglobulin A deficiency (sIgAD) is classified as a primary immunodeficiency and is diagnosed in individuals over the age of four who have serum IgA levels below 7 mg/dL, while maintaining normal IgG and IgM levels, adequate vaccine responses, and no evidence of secondary causes of hypogammaglobulinemia or T-cell immunodeficiencies.[1,2]

Despite being the most frequently encountered primary immunodeficiency, the symptomatic form of sIgAD is relatively rare, which limits the number of large-scale studies. Most individuals with sIgAD (approximately 85–90%) remain asymptomatic. However, when symptoms are present, they typically include recurrent sinopulmonary and gastrointestinal infections, allergic diseases, autoimmune conditions, and, less commonly malignancies.[1,3] Among symptomatic patients, recurrent upper and lower respiratory tract infections are the most common clinical presentation. These are usually mild to moderate in severity and viral in origin, whereas bacterial infections occur less frequently.[3]

The clinical manifestations of sIgAD vary widely, ranging from completely asymptomatic cases to individuals with significant immune-related complications. Some patients may exhibit allergic symptoms, frequent infections, or autoimmune features, and a subset may share phenotypic similarities with common variable immunodeficiency (CVID), suggesting a higher risk for progression to more severe immunodeficiency states.[4,5]

This broad clinical heterogeneity and variability in immune profiles highlight the importance of individualized approaches to diagnosis, follow-up, and treatment in patients with sIgAD. As such, managing sIgAD remains a clinical challenge for both healthcare providers and researchers.[6]

Although sIgAD is the most common form of primary immunodeficiency, large-scale cohort studies are limited due to the relatively low frequency of symptomatic cases. This makes sIgAD a significant challenge for both clinicians and researchers.[6]

Considering the wide variability in clinical phenotypes and the associated immunological differences among patients with IgA deficiency, it is crucial to establish appropriate strategies for their management, treatment, and follow-up.[6] In this context, our study aimed to provide a comprehensive evaluation of patients with sIgAD by analyzing demographic characteristics, infection frequency, associated autoimmune and allergic diseases, vaccine responses, and immunological findings such as IgG subclass and lymphocyte subset abnormalities.

Materials and Methods

This retrospective study included 45 patients diagnosed with sIgAD who were evaluated at the Pediatric Allergy and Immunology Departments of Şişli Hamidiye Etfal Training and Research Hospital by Professor Dr. Cemil Taşcıoğlu City Hospital between 2020 and 2025. IgAD was defined as a serum IgA level of less than 7 mg/dL in patients older than 4 years of age, with normal or elevated serum IgG and IgM levels, after the exclusion of other causes of hypogammaglobulinemia and T-cell immunodeficiencies.[7] Demographic, clinical, and laboratory data were collected through a review of patients' medical records, including sex, age at diagnosis, age at symptom onset, parental consanguinity, duration of breastfeeding, and follow-up information.

Laboratory data were obtained from routine clinical evaluations. Serum levels of immunoglobulin G, A, M, and E were measured in all patients. Additionally, IgG subclass levels were assessed by nephelometry in 22 patients, and lymphocyte subset analysis—including T, B, and NK cells—was performed in all 45 patients. Autoantibodies were evaluated in at least one sample in 40 patients, and specific antibody responses to vaccination were assessed in 44 patients. All laboratory values were interpreted according to age-specific reference ranges.[8,9]

Statistical analysis

Statistical analysis was conducted for all participants using the Jamovi 2.3.26 version (The Jamovi Project, Sydney, Australia). Descriptive statistics are reported according to the distributional characteristics of the data; mean and standard deviation (SD) were used for normally distributed variables, while median and interquartile range (IQR) were used for non-normally distributed variables. The normality of the data distribution was assessed using, for example, the Shapiro–Wilk test.

The figure was created by inputting data into GraphPad Prism 8XML version (GraphPad Software Inc., San Diego, California) and Adobe Illustrator 25.2.1 (Adobe Inc., San Jose, California).

Results

A total of 45 patients (20 females and 25 males) diagnosed with sIgAD and followed at two different centers were included in the study. The demographic and clinical characteristics of the patients are presented in Table 1.

Evaluation of the diagnostic process revealed that the most common reason for referral was recurrent upper respiratory tract infections (URTIs) (n = 7). Additionally, five patients presented with allergic symptoms, four with both recurrent URTIs and allergic symptoms, three with lower respiratory tract infections, two with lymphadenopathy, four with gastrointestinal symptoms, and two with herpes infections. One patient was referred for Hashimoto's thyroiditis, one for vitiligo, and one for recurrent otitis. In 10 patients, immunological evaluation was initiated due to frequent asthma exacerbations requiring beta-agonist use. Furthermore, two patients were evaluated during routine pediatric follow-up, two due to recurrent urinary tract infections, and one during hospitalization for infection (Table 2).

Assessment of annual infection frequencies revealed a median URTI frequency of 6.0 episodes per year (range: 0–12), pneumonia 0.0 (0–5), otitis 0.0 (0–4), meningitis 0.0 (0–0), and viral infections 6.0 (0–12). Frequent infection was defined according to previously published criteria as ≥8 episodes of upper respiratory tract infection

Table 1 Demographic and clinical characteristics of patients with sIgAD.

Gender, n (%)	
Female	20 (44.4%)
Male	25 (55.6%)
Current age of the patients (months) Median (range)	102 (48–204)
Age at symptom onset (months) Median (range)	24 (1–186)
Age at referral to the immunology clinic (month) Median (range)	88 (6–199)
Age at diagnosis (months) Median (range)	87 (48–192)
Follow-up period (months) Median (range)	12 (3–72)
Parental consanguinity, n (%)	8 (17.8%)
Allergic disorders, n (%)	30 (66.7%)
Allergic rhinitis	21 (46.7%)
Asthma	19 (42.2%)
Atopic dermatitis	16 (35.6%)
Chronic urticaria	7 (15.6%)
Aeroallergen sensitizations, n (%)	18 (41.9%)
House dust mites	
Dermatophagoides farinae	8 (18.6%)
Dermatophagoides pteronyssinus	8 (18.6%)
Pollens	
Weed pollen mix	2 (4.6%)
Plantago lanceolata (narrowleaf plantain)	1 (2.3%)
Artemisia vulgaris (mugwort)	1 (2.3%)
Grass pollen mix	1 (2.3%)
Tree pollen mix	1 (2.3%)
Olea europaea (olive tree)	1 (2.3%)
Molds	
Alternia tenuis	1 (2.3%)
Aspergillus fumigatus	1 (2.3%)
Mold mix	2 (4.6%)
Animal Dander	
Cat dander/epithelium	6 (13.9%)
Dog dander/epithelium	1 (2.3%)
Cockroach (*Blatella germinica*)	1 (2.3%)

n: Number of patients; sIgAD: Selective immunoglobulin A deficiency.

Table 2 Distribution of sIgAD patients according to clinical presentation at the time of diagnosis.

Reason for immunological evaluation	Number of patients (n)
Frequent beta-agonist use (asthma exacerbations)	10
Recurrent URTI	7
Allergic symptoms	5
Recurrent URTI + allergic symptoms	4
Gastrointestinal symptoms	4
Lower respiratory tract infections	3
Lymphadenopathy	2
Herpes infections	2
Routine pediatric check-up	2
Recurrent urinary tract infections	2
Vitiligo	1
Hashimoto's thyroiditis	1
Otitis media	1
Evaluation during hospitalization for infection	1

sIgAD: Selective immunoglobulin A deficiency; URTI: Upper respiratory tract infection.

per year or ≥2 episodes of pneumonia per year, in line with the warning signs of the Jeffrey Modell Foundation.[10] Due to frequent infections, four patients were on antibiotic prophylaxis, and one patient was receiving immunoglobulin replacement therapy (IgRT). One patient was prescribed acyclovir due to recurrent herpetic lesions, while the remaining three received amoxicillin, azithromycin, or trimethoprim-sulfamethoxazole.

Thirty patients (66.7%) had a history of at least one allergic disease. Allergic rhinitis was reported in 21 (46.7%) patients, asthma in 19 (42.2%), atopic dermatitis in 16 (35.6%), and chronic urticaria in 7 (15.6%). Among the 43 patients who underwent skin prick testing and/or specific IgE testing, aeroallergen sensitization was detected in 18 patients (41.9%). A family history of atopy was present in 17 patients, and a family history of autoimmune disease was reported in 4.

All 45 patients had serum IgG and IgM levels within age-appropriate reference ranges. Total IgE levels were elevated (>200 IU/mL) in 12 (26.7%) patients. Among 22 patients evaluated for IgG subclasses, IgG1 was normal in 16 (72.7%), low in 1 (4.5%), and elevated in 5 (22.7%); IgG2 was normal in 20 (90.9%) and elevated in 2 (9.1%); IgG3 was normal in 17 (77.3%), low in 4 (18.2%), and elevated in 1 (4.5%); and IgG4 was normal in 10 (45.5%), low in 5 (22.7%), and elevated in 7 (31.8%) (Figure 1A).

Vaccination history revealed that only one patient's family was vaccine-hesitant, and this patient was excluded from the evaluation of vaccine responses. All other 44 patients had received vaccinations according to the National Immunization Program of the Ministry of Health. No adverse reaction after vaccination was reported. Among the patients evaluated for vaccine-specific antibody responses, protective titers were detected in 31/43 (73%) for anti-HBs, 12/19 (63%) for varicella IgG, 16/19 (84%) for mumps IgG, 36/41 (88%) for measles IgG, and 19/20 (95%) for rubella IgG (Figure 1B).

Seven patients had a history of autoimmune disease, including Hashimoto's thyroiditis (n = 3), vitiligo (n = 2), immune thrombocytopenic purpura (n = 1), psoriasis (n = 1), and autoimmune hemolytic anemia (n = 1). Additionally, two patients had coexisting Familial Mediterranean Fever (FMF). Antithyroglobulin antibody was positive in 1 of 30 patients, and anti-TPO was positive in 3 of 40. Tissue transglutaminase IgA and IgG were tested in 40 patients, with no positive results. Antinuclear antibodies (ANA) were positive in 4 of 23 patients, while anti-dsDNA was negative in all patients tested (n = 23).

Lymphocyte subset analysis was performed simultaneously with complete blood counts in all patients.

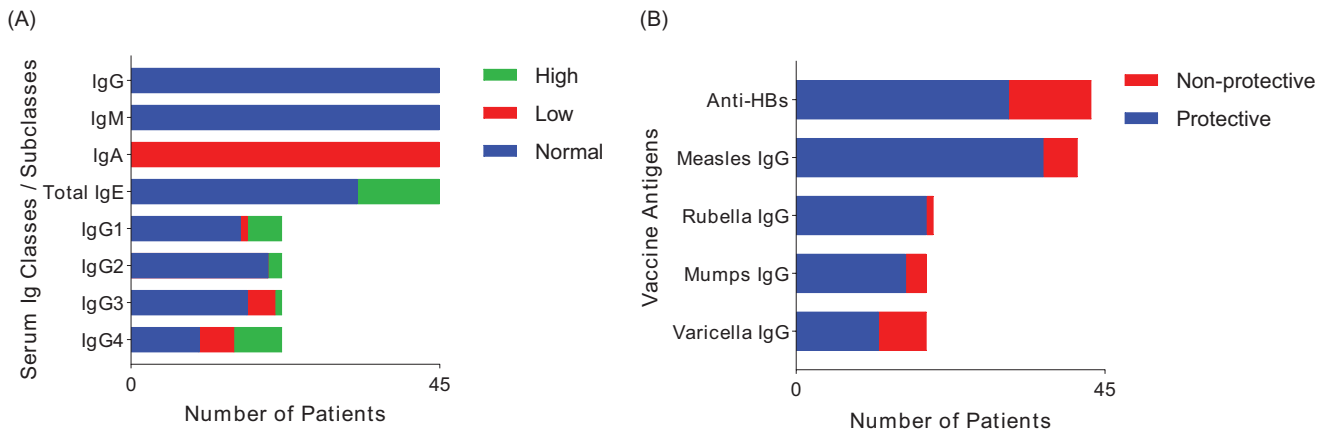

Figure 1 (A) Distribution of serum immunoglobulin (Ig) classes and subclasses among patients with selective IgA deficiency (sIgAD). Bars represent the number of patients with normal (blue), low (red), and high (green) levels for each immunoglobulin. Total IgE levels were elevated in a substantial number of patients, whereas IgA was low in all patients by definition. Minor variations were observed in IgG subclasses, particularly in IgG2 and IgG3. No cases of elevated IgA or IgM were detected. (B) Serologic IgG responses to vaccine antigens (hepatitis B surface antigen, measles, rubella, mumps, and varicella) in patients with selective IgA deficiency (sIgAD) are shown. Bars show the number of patients with protective (blue) and nonprotective (red) IgG levels against various vaccine antigens, including hepatitis B surface antigen (anti-HBs), measles, rubella, mumps, and varicella. The highest proportion of nonprotective antibody levels was observed for anti-HBs and measles IgG, while lower rates of seronegativity were noted for rubella, mumps, and varicella.

Total lymphocyte counts were > 1500/mm³ in all cases, and lymphopenia was not detected. Median percentages were as follows: CD3+ cells 72.5% (58.0–88.0), CD4+ 40.78% (26.67–59.0), CD8+ 27.0% (15.0–45.0), CD19+ 15.0% (7.0–30.0), and CD16+56+ NK cells 10.44% (2.0–20.22). Corresponding median absolute counts were CD3+ 2158/mm³ (1345–3801), CD4+ 1290/mm³ (565–2361), CD8+ 688.5/mm³ (333–1382), CD19+ 420/mm³ (258–1382), and CD16+56+ NK cells 282/mm³ (76–546). All percentages and absolute counts were within age-specific reference ranges, and no significant abnormalities suggestive of cellular immunodeficiency were identified.

The association between IgG3 and IgG4 subclass levels and frequency of infection was evaluated. For this purpose, patients with low IgG3 or IgG4 levels were compared to those with normal or elevated levels. Annual frequencies of URTIs, pneumonia, otitis, and viral infections were analyzed separately, and intergroup comparisons were performed using the Mann-Whitney U test. When patients with low IgG3 levels (n = 4) were compared to those with normal or high levels (n = 18), no statistically significant differences were observed in the frequency of URTIs (P = 0.8976), pneumonia (P = 0.5681), otitis (P = 1.0000), or viral infections (P = 0.7296). Similarly, among patients with low IgG4 levels (n = 5) and those with normal or high levels (n = 17), no significant difference was found in the frequency of URTIs (P = 0.4531). For the other infection types, statistical analysis could not be performed due to limited data.

Discussion

Selective IgA deficiency is considered the most common primary immunodeficiency (PID) based on studies conducted in the general population or among healthy blood donors.[1] In an epidemiological study conducted in Turkey, the prevalence of IgAD among healthy school children was found to be 1 in 188 in 2011.[11] Despite being frequently encountered, sIgAD presents a highly variable clinical phenotype, from asymptomatic individuals to patients suffering from recurrent infections, allergic diseases, autoimmune disorders, and even progression to CVID in a subset of cases.[6]

In our study of 45 pediatric patients followed at two tertiary care centers, recurrent infections—particularly URTIs—were the most common presenting symptom. This aligns with earlier reports identifying URTIs as a characteristic feature in symptomatic sIgAD.[4,12] Although infection was a frequent reason for initial evaluation, the overall annual infection burden was mild, and only a minority required long-term antibiotic prophylaxis or immunoglobulin replacement therapy, supporting the notion of a generally favorable course in childhood-onset sIgAD.

Although it is widely accepted that allergic diseases are more common in patients with sIgAD, the true prevalence remains uncertain, as studies from different countries have reported inconsistent findings, with some showing rates comparable to the general population. Therefore, it suggests that the prevalence varies depending on the ethnic background.[13] Nevertheless, the association between sIgAD and allergic diseases has been questioned by some researchers, and it has remained a subject of medical debate for more than five decades.[4,13–15] For example, Buckley et al. reported a rate of atopy as high as 58% among children and adults with sIgAD.[16] In contrast, Edwards et al. found an allergy or asthma prevalence of 13%, noting a higher frequency in younger individuals.[17] In a more recent study, Aytekin et al. reported allergic manifestations—including asthma, atopic dermatitis, allergic rhinitis or conjunctivitis, urticaria, drug allergy, and food allergy—in

43.2% of sIgAD patients aged 4–18 years.[12] In the study by Erkoçoğlu et al., 34.6% of patients had asthma, 27.2% had allergic rhinitis, and 11.1% had eczema, with positive skin prick test in 22.2%.[18] In comparison, a recent Turkish population-based study reported physician-diagnosed asthma, allergic rhinitis, and eczema in school-aged children at rates of 10.7, 16.9, and 2.6%, respectively.[19] These findings suggest a notably higher prevalence of asthma and eczema among children with sIgAD. In our study, allergic manifestations were observed in 66.7% of patients, with allergic rhinitis being the most common. This aligns with previous reports indicating increased rates of atopy in sIgAD, likely due to underlying mucosal immune dysregulation.[17,20] And aeroallergen sensitization was identified in 41.9% of the patients who underwent testing, highlighting a substantial burden of atopy among individuals with sIgAD. This finding reinforces the well-established association between sIgAD and allergic sensitization. Recent studies have increasingly emphasized the clinical relevance of routine atopic evaluation in patients with sIgAD, given that allergic manifestations—such as rhinitis, asthma, or eczema—may constitute the predominant clinical presentation in a significant proportion of cases. Accordingly, early recognition and management of allergic symptoms may play a crucial role in improving the quality of life in this patient population.

Autoimmune conditions, including Hashimoto's thyroiditis, vitiligo, and immune thrombocytopenic purpura, were documented in 13.3% (n = 6) of our patients. The prevalence of autoimmune disorders has been estimated to be 3–5 % in Western countries.[20,21] However, the prevalence of autoimmune disorders in patients with IgAD is highly variable. This rate is consistent with earlier reports indicating a 7–36% prevalence of autoimmune disorders in sIgAD.[14,18,20,21] The underlying mechanisms are still under investigation, but may include immune dysregulation, impaired clearance of immune complexes, and shared genetic susceptibility with other primary immunodeficiencies. Our findings support the need for longitudinal monitoring of autoimmunity in sIgAD, especially as some autoimmune manifestations may develop over time.[12,22,23]

Some individuals exhibited reduced IgG3 or IgG4 levels; however, there were no statistically significant differences in infection rates between patients with low versus normal or high subclass concentrations. The small number of patients with low subclass levels limits the statistical power of this analysis, and further studies with larger sample sizes are warranted to confirm these results.

While earlier studies suggested that concurrent IgG subclass deficiencies, particularly IgG2 or IgG3, in sIgAD patients heighten susceptibility to infection, recent research, including our findings, casts doubt on this association. The current evidence indicates that isolated IgG subclass deficiencies do not necessarily serve as independent predictors of infection risk. Consequently, the routine measurement of IgG subclasses in sIgAD remains controversial and may offer limited additional clinical utility.[24–26]

Vaccine-specific antibody responses were assessed in our study and found to be largely adequate, with high seroprotection rates against hepatitis B, measles, mumps, rubella, and varicella. Although impaired specific antibody responses have been reported in some sIgAD patients, most individuals in our study demonstrated preserved functional humoral immunity. This supports previous work indicating that vaccine responses are generally reliable in sIgAD unless additional immune defects coexist.[17]

Overall, our findings reinforce the heterogeneity of clinical presentations in sIgAD. The absence of a clear association between IgG subclass deficiencies and infection rates in our study highlights the importance of individualized risk assessment. This study has several limitations, including its retrospective design, the presence of missing data, a relatively short follow-up, and a limited sample size, which reduce the statistical power of some subgroup analyses. Nevertheless, the comprehensive evaluation of autoimmune and allergic manifestations, infection frequency, vaccine responses, and lymphocyte subset abnormalities represents an important strength of our work. Taken together, these findings provide valuable insights into the clinical spectrum of sIgAD in our setting. As highlighted in the Abstract, multicenter and longitudinal prospective studies with larger patient cohorts and standardized criteria are warranted to better define long-term outcomes and to guide tailored management strategies.

Conclusion

Selective IgA deficiency represents a heterogeneous clinical entity with varying presentations ranging from asymptomatic cases to patients with significant allergic, infectious, or autoimmune complications. In our pediatric cohort, the majority of patients exhibited mild disease courses, with a high prevalence of allergic manifestations and a smaller subset showing autoimmune comorbidities. Despite the presence of IgG3 and IgG4 subclass deficiencies in some patients, no significant association with infection burden was observed. Vaccine responses were generally preserved, indicating adequate functional humoral immunity in most cases. These results emphasize the need for individualized clinical follow-up rather than routine subclass testing and support continued research through prospective, multicenter studies to optimize diagnostic and therapeutic strategies in sIgAD.

Statement of Ethics

This study protocol was reviewed and approved by the Clinical Research Ethics Committee of the Şişli Hamidiye Etfal Training and Research Hospital Health Practice and Research Center (SUAM) on January 14, 2025 (Decision No: 2891). Written informed consent was obtained from the parents or legal guardians of all participants included in the study.

Data Availability Statement

The data that support the findings of this study are not publicly available due to institutional and ethical restrictions, but can be shared by the corresponding author upon reasonable request.

Author's Contributions

E.Y.G. and S.S.C. conceptualized and supervised the study. E.Y.G., N.C., B.O., I.K.K., D.O., and S.S.C. provided patient care, collected clinical data, and conducted thequestionnaires. Calculations and statistical analyses were performed by E.Y.G. and E.Y.G., and S.S.C. wrote the paper. All authors reviewed and approved the final version of the manuscript.

Conflicts of Interest

The authors have no conflicts of interest to declare.

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