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Clinical presentation of selective IgA deficiency in paediatric patients: a single-centre retrospective study

Obraz kliniczny selektywnego niedoboru IgA u dzieci – retrospektywne badanie jednoośrodkowe

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Abstract

Introduction: Selective immunoglobulin A deficiency is the most common primary immunodeficiency. It is defined as a serum immunoglobulin A concentration <7 mg/dL in individuals older than four years, with normal immunoglobulin G and immunoglobulin M levels. Its clinical presentation is heterogeneous, ranging from asymptomatic cases to recurrent infections, allergic diseases, and autoimmune disorders. Data regarding the Polish paediatric population remain limited. **Objective:** To characterise the clinical and immunological features of children with selective immunoglobulin A deficiency hospitalised in a single centre, with particular emphasis on the frequency of recurrent infections, allergic manifestations, and selected immunological parameters. **Materials and methods:** A retrospective analysis of medical records from 3,063 patients hospitalised in a paediatric department was performed. Selective immunoglobulin A deficiency was identified in 22 children aged 4–14 years (median age 6 years; 11 boys and 11 girls). Clinical data and serum concentrations of immunoglobulin G, immunoglobulin M, immunoglobulin E, and complement components (C3, C4) were evaluated. **Results:** Clinical symptoms were present in 20/22 patients (90.9%). Recurrent infections and allergic manifestations were equally prevalent (14/22; 63.6%), and upper respiratory tract infections occurred in 10/22 (45.5%). Immunoglobulin M deficiency was found in 4/22 (18.2%), and elevated immunoglobulin E levels in 6/22 (27.3%). Serum immunoglobulin G concentrations were significantly higher in children with selective immunoglobulin A deficiency compared with the reference group [median 1,290 (IQR 1,1140–1,482) vs. 1,020 (IQR 872–1,180) mg/dL; $p = 0.004$]. **Conclusions:** The phenotype of selective immunoglobulin A deficiency in the studied children was characterised by the coexistence of recurrent respiratory tract infections and allergic manifestations. These findings suggest the presence of complex disturbances of mucosal immunity and immune dysregulation in the course of this condition.

Keywords: immunoglobulins, primary immunodeficiency, allergy, respiratory tract infections, selective immunoglobulin A deficiency

Streszczenie

Wprowadzenie: Selektywny niedobór immunoglobuliny A jest najczęstszym pierwotnym niedoborem odporności. Definiuje się go jako stężenie immunoglobuliny A <7 mg/dl u osób powyżej czwartego roku życia, przy prawidłowych stężeniach immunoglobuliny G i immunoglobuliny M. Obraz kliniczny jest zróżnicowany – od przebiegu bezobjawowego po nawracające zakażenia, choroby alergiczne i autoimmunologiczne. Dane dotyczące polskiej populacji pediatrycznej są ograniczone. **Cel:** Charakterystyka kliniczna i immunologiczna dzieci z selektywnym niedoborem immunoglobuliny A hospitalizowanych w jednym ośrodku, ze szczególnym uwzględnieniem częstości nawracających zakażeń, manifestacji alergicznych oraz wybranych parametrów immunologicznych. **Materiał i metody:** Retrospektywna analiza dokumentacji medycznej 3063 pacjentów hospitalizowanych na oddziale pediatrycznym, spośród których u 22 dzieci w wieku 4–14 lat (mediana 6 lat; 11 chłopców, 11 dziewczynek) stwierdzono selektywny niedobór immunoglobuliny A. Oceniono dane kliniczne oraz stężenia immunoglobuliny G, immunoglobuliny M, immunoglobuliny E i składowych dopełniacza (C3, C4). **Wyniki:** Objawy kliniczne

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wystąpiły u 20/22 pacjentów (90,9%). Nawracające infekcje i manifestacje alergiczne stwierdzono u 14/22 (63,6%), a zakażenia górnych dróg oddechowych u 10/22 (45,5%). Niedobór immunoglobuliny M występował u 4/22 (18,2%), a podwyższone stężenie immunoglobuliny E u 6/22 (27,3%). Stężenie immunoglobuliny G było istotnie statystycznie wyższe u dzieci z selektywnym niedoborem immunoglobuliny A niż w grupie referencyjnej [mediana: 1290 (IQR 1140–1482) *vs* 1020 (IQR 872–1180) mg/dl; $p = 0{,}004$]. **Wnioski:** Fenotyp selektywnego niedoboru immunoglobuliny A u badanych dzieci charakteryzował się współwystępowaniem nawracających zakażeń układu oddechowego oraz objawów alergicznych. Obserwacje te wskazują na obecność złożonych zaburzeń odporności śluzówkowej i dysregulacji immunologicznej w przebiegu tego schorzenia.

Słowa kluczowe: immunoglobuliny, pierwotny niedobór odporności, alergia, zakażenia układu oddechowego, selektywny niedobór immunoglobuliny A

INTRODUCTION

Selective immunoglobulin A deficiency (SIgAD) is the most common primary humoral immunodeficiency in Caucasian populations, with an estimated prevalence of approximately 1:600[1,2]. According to the criteria of the European Society for Immunodeficiencies (ESID), the diagnosis requires a serum immunoglobulin A (IgA) level below 7 mg/dL (0.07 g/L) with normal immunoglobulin G (IgG) and immunoglobulin M (IgM) concentrations in individuals aged 4 years or older, after exclusion of secondary causes of hypogammaglobulinaemia and T-cell defects[3]. The lower age limit reflects the physiological delay in IgA maturation; longitudinal studies indicate that a substantial proportion of children identified with SIgAD before adolescence may subsequently normalise their IgA levels[4].

IgA is the predominant immunoglobulin class at mucosal surfaces, contributing to pathogen exclusion, regulation of commensal microbiota, and modulation of allergic sensitisation[5,6]. In SIgAD, the absence of both secretory and serum IgA compromises mucosal barrier integrity, potentially predisposing affected individuals to recurrent respiratory and gastrointestinal infections and increasing exposure to aeroallergens and food antigens[7]. The clinical spectrum of SIgAD is heterogeneous, encompassing recurrent infections, atopic manifestations, autoimmune conditions, and – in a minority of cases – progression to common variable immunodeficiency (CVID)[8,9]. Importantly, a considerable proportion of individuals with SIgAD remain entirely asymptomatic throughout life[1], underscoring the difficulty of estimating true disease burden from clinic-based cohorts.

Published studies report wide variation in the prevalence of symptoms[2,10,11]. This heterogeneity reflects methodological differences, including study design, patient selection (symptomatic vs. unselected), age range, ethnic background, and diagnostic criteria. In the largest paediatric series, recurrent respiratory infections are consistently the most frequent presenting feature, with atopic manifestations constituting the second most common clinical phenotype[12,13]. Data on SIgAD in the Polish paediatric population remain limited. The present study describes the clinical and immunological characteristics of children with SIgAD identified at a single paediatric ward.

MATERIALS AND METHODS

Study design and patient selection

This retrospective study analysed medical records of children admitted to the Department of Paediatrics, Paediatric Nephrology and Allergology, Military Institute of Medicine – National Research Institute, Warsaw, Poland, between January 2022 and December 2024. Of 3,063 patients in whom immunoglobulin levels were measured during this period, 22 met the inclusion criteria for the SIgAD study group. Eligible patients were aged 4 years or older at the time of assessment and had a serum IgA level below 7 mg/dL with normal serum IgG and IgM concentrations, in accordance with ESID criteria[3]. Patients were excluded if a secondary aetiology of hypogammaglobulinaemia was identified, including drug-induced immunosuppression or viral infections (e.g. cytomegalovirus, Epstein–Barr virus).

For the purpose of comparing serum IgG concentrations, an age-matched reference group was selected from children hospitalised during the same period who had normal serum IgA levels ($n = 1{,}814$). Children with known immunodeficiency, haematological malignancy, or immunosuppressive therapy were excluded from the reference group.

The study was conducted in accordance with the Declaration of Helsinki. Under applicable local regulations, formal ethics committee approval was not required for this retrospective record-based analysis.

Data collection

The following variables were extracted from medical records: age, sex, serum IgA (mg/dL), IgG (mg/dL), IgM (mg/dL), IgE (IU/mL), complement components C3 and C4 (mg/dL), and serum alpha-1-antitrypsin (AAT) concentration.

Immunoglobulin levels were classified as deficient or elevated according to age-adjusted reference ranges. Concomitant clinical diagnoses included recurrent upper respiratory tract infections (URTI; defined as ≥8 episodes per year), recurrent lower respiratory tract infections (LRTI; ≥2 episodes per year), recurrent urinary tract infections (UTI; ≥3 episodes per year or ≥2 in the past six months), chronic rhinosinusitis (symptoms lasting

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≥12 weeks), recurrent acute otitis media (≥3 episodes per six months or ≥4 per year), skin disorders, adenoidal hypertrophy, bronchial asthma, autoimmune diseases, inhalant allergy, food allergy, intestinal parasitosis, hepatic manifestations, chronic gastrointestinal symptoms, coeliac disease, and cystic fibrosis[3]. All comorbidities were recorded as binary variables (present/absent) based on clinician-established diagnoses documented in the medical records.

Statistical analysis

Statistical analyses were performed using Python 3.12 with the SciPy library (v1.11). Normality of continuous variables was assessed using the Shapiro–Wilk test. Where normality was not confirmed, non-parametric tests were applied. Continuous variables are reported as median with interquartile range (IQR). Comparisons between two groups were performed using the Mann–Whitney *U* test for non-normally distributed data. A p-value < 0.05 was considered statistically significant.

RESULTS

Patient characteristics

Of the 3,063 patients in whom immunoglobulin levels were measured during the study period, 22 children (0.72%) met the inclusion criteria for SIgAD. The final cohort comprised 11 males (50%) and 11 females (50%). The median age was 6 years (IQR 4–8; range 4–14 years).

Median serum IgG was 1,290 mg/dL (IQR 1,140–1,482; n = 20; two patients lacked IgG measurement). Median IgM was 100 mg/dL (IQR 62–115), and median IgE was 37 IU/mL (IQR 12–66; n = 21). Concomitant IgM deficiency was present in 4/22 patients (18.2%; individual IgM values: 38, 44, 45, and 59 mg/dL; patient ages: 4, 7, 9, and 13 years). Elevated IgE above the age-adjusted reference range was documented in 6/22 patients (27.3%). One patient (4.5%) had complement C3 deficiency. Demographic and immunological characteristics are summarised in Tab. 1.

Variable	Value
Total patients	22
Male/Female, *n* [%]	11 (50%)/11 (50%)
Age, median (IQR), years	6 (4–8); range 4–14
Age 4–6 years, *n* [%]	14 (63.6%)
Age 7–10 years, *n* [%]	4 (18.2%)
Age >10 years, *n* [%]	4 (18.2%)
Serum IgA	<7 mg/dL (all patients)
Serum IgG, median (IQR) [mg/dL]	1,290 (1,140–1,482); $n = 20$
Serum IgM, median (IQR) [mg/dL]	100 (62–115)
Serum IgE, median (IQR) [IU/mL]	37 (12–66); $n = 21$
IgM deficiency, *n* [%]	4 (18.2%)
Elevated IgE (above age-adjusted normal), *n* [%]	6 (27.3%)
C3 deficiency, *n* [%]	1 (4.5%)
IQR – interquartile range.	

*Tab. 1. Demographic and immunological characteristics of the study cohort (*n = *22)*

Clinical manifestations

Overall, 20 of 22 patients (90.9%) presented with at least one clinical manifestation. Recurrent infections and allergic or immune-mediated skin manifestations were equally prevalent, each occurring in 14/22 patients (63.6%). Both phenotypes coexisted in 8 patients (36.4%); 6 patients had exclusively infectious manifestations, 6 had exclusively allergic or dermatological manifestations, and only 2 patients (9.1%) had no recorded comorbidity.

Recurrent URTI were the most frequently documented infectious manifestation (10/22, 45.5%), followed by recurrent LRTI (6/22, 27.3%). Recurrent acute otitis media was present in 3 patients (13.6%) and chronic rhinosinusitis in 1 (4.5%). Adenoidal hypertrophy was identified in 8/22 patients (36.4%); 6 of whom also had recurrent URTI.

Skin manifestations were present in 8/22 children (36.4%), comprising atopic dermatitis in 4 patients (18.2%), urticaria in 3 (13.6%), and psoriasis in 1 (4.5%). Inhalant allergy was documented in 6 patients (27.3%) and food allergy in 2 (9.1%). Bronchial asthma was identified in a single patient (4.5%).

Coeliac disease was diagnosed in 2 patients (9.1%). Two patients (9.1%) had a pre-existing diagnosis of Alport syndrome. No cases of cystic fibrosis, intestinal parasitosis, hepatic manifestations, or chronic gastrointestinal symptoms were recorded.

The full distribution of clinical features is presented in Tab. 2.

Immunological findings

Patients with elevated IgE were younger than those with normal IgE (median age 4 years (IQR 4–5) vs. 6 years (IQR 5–9); Mann–Whitney U, $p = 0.016$, rank-biserial $r = 0.58$). All six patients with elevated IgE were aged 6 years or younger.

Concomitant IgM deficiency was present in 4/22 patients (18.2%; IgM values: 38, 44, 45, and 59 mg/dL). No sex-based differences were observed for any clinical or immunological variable.

Children with SIgAD had significantly higher serum IgG concentrations than the age-matched reference group with normal IgA levels [median: 1290 (IQR 1,140–1,482) vs. 1,020 (IQR 872–1,180) mg/dL; Mann–Whitney U, $p = 0.004$, rank-biserial $r = 0.41$].

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Clinical feature	*n*	%
Any recurrent infection	14	63.6
Recurrent URTI	10	45.5
Recurrent LRTI	6	27.3
Recurrent acute otitis media	3	13.6
Chronic rhinosinusitis	1	4.5
Recurrent UTI	1	4.5
Adenoidal hypertrophy	8	36.4
Skin disease	8	36.4
• Atopic dermatitis	4	18.2
• Urticaria	3	13.6
• Psoriasis	1	4.5
Inhalant allergy	6	27.3
Food allergy	2	9.1
Asthma	1	4.5
Coeliac disease	2	9.1
Both infection and allergic/skin manifestation	8	36.4
Infection only	6	27.3
Allergic/skin manifestation only	6	27.3
Neither infection nor allergic/skin manifestation	2	9.1
LRTI – lower respiratory tract infection; **URTI** – upper respiratory tract infection; **UTI** – urinary tract infection.		

Tab. 2. Clinical manifestations in 22 children with SIgAD

RESULTS

This retrospective single-centre study characterises the clinical and immunological profile of 22 children aged 4–14 years with SIgAD. The principal findings were the high prevalence of symptomatic disease (90.9%) and the predominance of recurrent infections and allergic manifestations.

The frequency of recurrent infections (63.6%) is consistent with observations from larger European paediatric studies. Moschese et al., in a longitudinal Italian study of 103 symptomatic children, reported infectious manifestations in approximately 62% of patients[12]. Similarly, Lougaris et al. identified respiratory tract infections as the most common clinical presentation in a single-centre cohort of 184 patients[13]. Higher rates have also been described; Pavelescu et al. reported recurrent infections in 86% of children in a Romanian hospital-based cohort restricted to symptomatic, hospitalised patients[14]. These discrepancies likely reflect differences in study design, referral patterns, diagnostic criteria, and healthcare systems. As the present cohort was identified from a general paediatric database without selection based on symptom severity, it may represent a broader clinical spectrum than studies limited to symptomatic populations, although referral bias related to paediatric allergology care cannot be excluded.

The prevalence of allergic manifestations (63.6%) was at the upper end of the range reported in the literature (13–84%)[2,10,11]. Comparisons between studies remain challenging because of substantial methodological heterogeneity, including differences in diagnostic criteria, age distribution, ethnic background, and the distinction between sensitisation and clinically confirmed allergy. A similar prevalence (62.7%) was reported by Ulaș et al. in a Turkish paediatric cohort of 110 patients[7], whereas lower frequencies have been described in other European studies[12,15]. These discrepancies highlight the limited comparability of clinic-based cohorts.

Cutaneous manifestations were particularly common in the present series (36.4%), predominantly atopic dermatitis (18.2%) and urticaria (13.6%). Psoriasis was identified in one patient (4.5%). Reported rates of skin involvement in SIgAD vary widely (4.6–57.8%), likely owing to differences in population characteristics and diagnostic definitions[2,7].

Bronchial asthma was documented in only one patient (4.5%), markedly lower than the prevalence reported in some studies (35–58%)[2,7]. This discrepancy may partly reflect the young age of our cohort, as 63.6% of patients were aged 4–6 years – a period during which a definitive diagnosis of asthma is often difficult and may overlap with recurrent preschool wheezing.

Adenoidal hypertrophy was observed in 8/22 patients (36.4%). This finding should be interpreted in the context of the cohort's age structure, as adenoidal enlargement is a common physiological phenomenon in early childhood, with peak prevalence between 3 and 8 years of age[16]. The predominance of children aged 4–6 years in the present study likely contributed substantially to the observed frequency. Chronic antigenic stimulation related to recurrent viral and bacterial respiratory infections is also a recognised driver of adenoidal hyperplasia[16]. In this cohort, recurrent URTI coexisted in 6 of 8 children with adenoidal hypertrophy, suggesting that lymphoid enlargement was primarily secondary to persistent mucosal infectious stimulation. Although prominent lymphoid hyperplasia has been described in primary immunodeficiencies involving impaired B-cell class switching, including SIgAD, the direct pathogenic relationship remains unclear. Consequently, the high prevalence of adenoidal hypertrophy in our cohort is more plausibly explained by recurrent infections and young age than by a disease-specific manifestation of SIgAD.

Autoimmune diseases are well-recognised comorbidities in SIgAD, reported in 7–36% of patients, most commonly including coeliac disease, autoimmune thyroid disease, systemic lupus erythematosus, rheumatoid arthritis, and inflammatory bowel disease[1,7]. The underlying pathogenesis is thought to involve impaired mucosal immune regulation due to the absence of secretory IgA, resulting in increased epithelial penetration of luminal antigens, persistent antigenic stimulation, and defective maintenance of immune tolerance. Shared genetic predisposition,

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particularly involving HLA-DQ alleles within the major histocompatibility complex (MHC), may additionally contribute to the association between SIgAD and autoimmunity[1,9]. In the studied cohort, coeliac disease was diagnosed in 2/22 patients (9.1%), consistent with previously reported paediatric prevalence rates of 2–14%[7,12]. Importantly, conventional tTG-IgA-based screening has limited diagnostic utility in SIgAD and should be replaced by IgG-based serological assays or intestinal biopsy[17]. Psoriasis was identified in one patient and may represent an additional manifestation of immune dysregulation, despite its predominantly IL-17/IL-23-driven inflammatory mechanism.

No autoimmune disorders other than coeliac disease were identified in our cohort. However, this finding should be interpreted with caution, as many autoimmune conditions associated with SIgAD typically manifest later in adolescence or adulthood[1,18,19]. Given the young age of the study population (median 6 years), the absence of systemic autoimmune disease most likely reflects limited age-dependent clinical expression rather than a genuinely lower predisposition. Longitudinal follow-up will therefore be required to assess the cumulative risk of autoimmunity in this group.

Two patients in our cohort had a pre-existing diagnosis of Alport syndrome. As Alport syndrome results from mutations in the *COL4A3*, *COL4A4*, or *COL4A5* genes on chromosomes 2 and X, whereas genetic susceptibility to SIgAD is primarily associated with loci within MHC region on chromosome 6[1,9], no shared pathogenic mechanism has been established. Their coexistence in the present cohort is therefore most likely incidental.

Two immunologically relevant findings merit emphasis. First, elevated IgE levels were significantly associated with younger age. All six patients with increased IgE concentrations were aged 6 years or younger, supporting the concept that atopic sensitisation in selective IgA deficiency (SIgAD) is an early and prominent feature of the disease course[2,20]. This observation is supported by known immunological mechanisms. Deficiency of secretory IgA at mucosal surfaces compromises antigen exclusion, facilitating allergen penetration and promoting IgE-mediated sensitisation[6,16]. In addition, reduced levels of serum monomeric IgA may impair inhibitory ITAMi signalling via the Fcα receptor (FcαRI), leading to enhanced immune activation and, consequently, increased IgE responses[21]. These mechanisms may be particularly relevant in early childhood, when the mucosal immune system remains functionally immature and physiological production of secretory IgA is still developing[4,6].

Secondly, children with SIgAD demonstrated significantly higher serum IgG concentrations than age-matched controls with normal IgA levels, and this association remained significant after adjusting for age. This finding is consistent with the recognised phenomenon of compensatory IgG upregulation in SIgAD, in which enhanced systemic IgG responses partially compensate for the absence of mucosal IgA[1,22]. Fadlallah et al. showed that patients with SIgAD develop augmented systemic IgG responses directed against commensal bacteria normally targeted by secretory IgA[23]. Despite this compensatory mechanism, IgG cannot fully replace the protective functions of secretory IgA, as it is transported across the mucosal epithelium far less efficiently[6,22]. As a result, mucosal immune defence remains impaired, likely contributing to the persistent susceptibility to respiratory and gastrointestinal infections observed in SIgAD despite elevated systemic IgG concentrations. Clinically, normal or increased serum IgG levels in children with SIgAD should therefore be interpreted not as evidence of preserved immune competence, but rather as a marker of ongoing immunological compensation.

Concomitant IgM deficiency was identified in 4/22 patients (18.2%). Previous studies suggest that combined IgA and IgM deficiency may be associated with a more severe infectious phenotype[1,9]; however, the limited number of affected patients in the studied cohort does not allow reliable assessment of potential clinical correlations.

Complement C3 deficiency was detected in one patient. Although complement abnormalities may contribute to increased susceptibility to recurrent bacterial infections[1], the isolated occurrence does not support a broader association with SIgAD.

Several limitations of this study should be acknowledged. First, the retrospective single-centre design and relatively small sample size (n = 22) limit statistical power and preclude robust multivariable analyses. Second, the study population is subject to referral bias, as patients evaluated in a paediatric allergology centre are more likely to present with symptomatic disease than the broader SIgAD population, which includes many asymptomatic individuals. Third, clinical data were derived from existing medical records using binary categorisation of diagnoses, which may have resulted in underreporting of milder or transient manifestations. Finally, no functional immunological assays, immunophenotyping, or cytokine profiling were performed, limiting the mechanistic interpretation of the observed immunological abnormalities. Further prospective, multicentre studies with larger cohorts and standardised diagnostic protocols are required to validate these observations.

CONCLUSIONS

Selective IgA deficiency in the studied paediatric cohort was associated with a high frequency of clinical manifestations, with symptoms present in more than 90% of patients. The disease phenotype was characterised by the coexistence of recurrent infections and allergic manifestations, supporting the concept of combined mucosal immune dysfunction and immune dysregulation in the course of this condition.

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Conflict of interest

The authors declare no conflicts of interest.

Author contribution

Original concept of study; writing of manuscript: KK, AB. Collection, recording and/or compilation of data: NG, KM, DG, AZ, SW. Analysis and interpretation of data: KK, NG, KM, AZ, AB. Critical review of manuscript: KK, AT, AR, AB. Final approval of manuscript: KK, AT, AB.

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