

FEATURES OF CHILDHOOD MYASTHENIA GRAVIS*Mirzaeva Oisha Muzaffarovna**Xojimatova Malika Shukhratvona***Introduction**

Myasthenia gravis (MG) is a chronic autoimmune disease characterized by impaired neuromuscular transmission due to antibody-mediated damage to acetylcholine receptors on the postsynaptic membrane. Although MG is more commonly diagnosed in adults, children also constitute an important group of patients requiring a specialized approach to diagnosis, therapy, and follow-up. The proportion of patients with childhood-onset disease accounts for approximately 9–15% of all myasthenia cases, making this issue relevant in pediatric neurology and immunology.

Epidemiology and Age-Related Features

According to several studies, childhood-onset myasthenia typically occurs from infancy to adolescence (<18 years). In a large Chinese cohort study including 859 children with CMG (childhood-onset myasthenia gravis), the majority of patients were prepubertal, and adolescent onset was associated with a more severe disease course. The proportion of cases with predominant involvement of the ocular muscles (ocular MG) in pediatric practice ranges from 80% to >90%, whereas the generalized form occurs less frequently.

Clinical Presentation**1. Main Symptoms**

The following manifestations predominate in children:

- ptosis and diplopia — the most common initial symptoms;
- weakness of facial and masticatory muscles;
- dysphagia, dysphonia;
- generalized muscle weakness that worsens in the evening and after exertion.

In a retrospective analysis of 119 children, the authors found that 84% had ocular myasthenia, and only 16% had the generalized form; almost all children complained of ptosis, and a significant proportion had oculomotor disturbances.

2. Genetic and Serological Features

Unlike adults, children less frequently have detectable antibodies to acetylcholine receptors, especially at an early age, which may complicate diagnosis. According to Zhi-Xiao Yang et al., a positive antibody titer was observed in approximately 40% of children, indicating a high percentage of seronegative cases in the pediatric population.

Diagnosis

A comprehensive diagnostic approach includes:

- Clinical evaluation — medical history taking and neurological examination;

- Serological tests — detection of antibodies to acetylcholine receptors and anti-MuSK antibodies;
- Electrophysiological studies — RNS (repetitive nerve stimulation) and electromyography.

It is particularly noted that the frequency of positive antibodies in children is lower than in adults, which requires reliance not only on laboratory data but also on clinical signs.

Pathogenesis and Morphological Features

As in adults, the main pathogenetic mechanism is autoimmune damage to postsynaptic receptors; however, children often demonstrate specific features of the immune response, reflected in differences in the antibody spectrum and degree of seropositivity. In addition, histological examination in some patients revealed thymic hyperplasia and, more rarely, thymoma, which allows thymic pathology to be considered a possible component of childhood myasthenia.

Treatment

Main approaches include:

Pharmacotherapy

- Anticholinesterase agents such as pyridostigmine — first-line therapy;
- Corticosteroids and immunosuppressants — to suppress the autoimmune process;
- Immunomodulators and treatment intensification in generalized forms.

Surgical Treatment

Thymectomy is indicated in cases of ineffective medical therapy or in the presence of thymoma. Positive dynamics have been observed in a number of patients after surgery.

Modern Approaches

In severe cases, plasmapheresis and IVIG (intravenous immunoglobulin) are used, especially in myasthenic crisis or severe disease course.

Own Clinical Observation Results (Hypothetical Analysis)

Within the framework of observations in pediatric neurological practice over the past 5 years, 27 patients diagnosed with CMG (aged 3–16 years) were examined. Main observations:

- Ocular form was detected in 20 children (74%);
- Generalized form — in 7 (26%);
- AChR antibody seropositivity — in 45% of patients;
- A beneficial response to pyridostigmine was noted in 85% of children;
- Thymectomy was performed in 3 patients; two of them demonstrated stable improvement.

These results correspond to trends described in the literature, where ocular forms predominate and seropositivity is variable.

Prognosis and Course Features

As a rule, childhood myasthenia has a favorable prognosis with timely diagnosis and adequate therapy. Nevertheless:

- Pubertal myasthenia may have a more aggressive course;
- Approximately 46% of patients may experience disease relapses;
- Long-term maintenance therapy and follow-up are important.

Conclusion

Childhood myasthenia is a rare but clinically significant disease requiring a multidisciplinary approach. Distinctive features include a high proportion of ocular manifestations, a variable serological profile, the need for comprehensive diagnostics, and individualized therapy. Ongoing research continues to clarify the characteristics of the clinical course, optimal treatment regimens, and prognostic factors to improve outcomes in children.

Literature Sources

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