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ALGORITHM FOR THE DIAGNOSIS AND MANAGEMENT OF PEDIATRIC MYASTHENIA GRAVIS

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Abstract

Myasthenia gravis in children is a rare autoimmune disorder of neuromuscular transmission characterized by pathological muscle fatigability and variable weakness of skeletal muscles. Despite its relative rarity in pediatric practice, this condition has significant clinical importance due to the risk of generalization, development of bulbar disorders, and myasthenic crises. Clinical manifestations in children are diverse and are often limited to oculomotor disturbances, which complicates early diagnosis. In addition, the pediatric population is characterized by a high frequency of seronegative forms, which reduces the diagnostic value of standard immunological markers and requires a comprehensive approach to diagnostic verification.

The article presents current data on the clinical and pathogenetic features of myasthenia gravis in children, as well as the results of analysis of our own clinical observations. The main clinical forms of the disease, diagnostic criteria, the capabilities of electrophysiological and immunological methods, and principles of medical and surgical treatment are discussed. Special attention is given to early detection and optimization of therapeutic strategies depending on the form and severity of myasthenia gravis. The obtained data indicate the predominance of oculomotor forms of myasthenia in children, the high effectiveness of anticholinesterase therapy, and the importance of timely administration of immunosuppressive agents in generalized forms of the disease. Early diagnosis and an individualized treatment approach improve prognosis and quality of life in pediatric patients.

Keywords: myasthenia gravis, children, neuromuscular transmission, autoimmune diseases, diagnosis, treatment.

Introduction

Myasthenia gravis belongs to the group of autoimmune disorders of neuromuscular transmission and is caused by the formation of antibodies against components of the postsynaptic membrane of the neuromuscular junction, leading to decreased efficiency of nerve impulse transmission and the development of pathological muscle fatigability. The disease is traditionally considered a condition of young and middle-aged adults; however, pediatric forms of myasthenia have independent clinical significance and require a special diagnostic and therapeutic approach. According to epidemiological studies, disease onset in childhood accounts for approximately 10–15% of all myasthenia cases. In pediatric practice, the disease often manifests as isolated ocular motor disturbances such as ptosis and diplopia, which may mimic ophthalmologic or neuro-ophthalmologic pathology and lead to delayed diagnosis.

A significant feature of pediatric myasthenia is the high frequency of seronegative variants, which limits the diagnostic value of standard immunological tests and increases the role of clinical assessment and electrophysiological methods.

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Diagnostic Algorithm

Stage	Method	Purpose	Comment
1	Clinical examination	Detection of fatigability, ptosis, diplopia, dysphagia	Symptoms worsen in the evening and decrease after rest
2	Anticholinesterase test	Confirmation of reversibility of weakness	Improvement within 30–60 minutes
3	Serology: AChR-Ab, MuSK-Ab	Confirmation of autoimmune origin	Seronegative forms are common in children
4	Electrophysiology (RNS, SFEMG)	Detection of decrement	Amplitude reduction ≥10%
5	Chest CT / MRI	Evaluation of thymus	Hyperplasia, rarely thymoma
6	Differential diagnosis	Exclusion of congenital myasthenic syndromes, myopathies, brain tumors	Mandatory if onset occurs before 2 years

Treatment Algorithm

Severity	Main Therapy	Additional Measures
Ocular form	Pyridostigmine	Follow-up every 3 months
Generalized	Pyridostigmine + glucocorticosteroids	Azathioprine if resistant
Severe course	Glucocorticosteroids + immunosuppressants	IVIG or plasmapheresis
Refractory form	Second-line immunosuppressants	Consider thymectomy
Myasthenic crisis	Mechanical ventilation + IVIG / plasmapheresis	Elimination of provoking factors

Clinical Study

A study was conducted involving 27 children diagnosed with myasthenia gravis at the Andijan Regional Multidisciplinary Children’s Hospital in the pediatric neurology departments. The mean age of disease onset was 7.4 ± 2.1 years. Among the patients, 74% had the ocular form and 26% had the generalized form.

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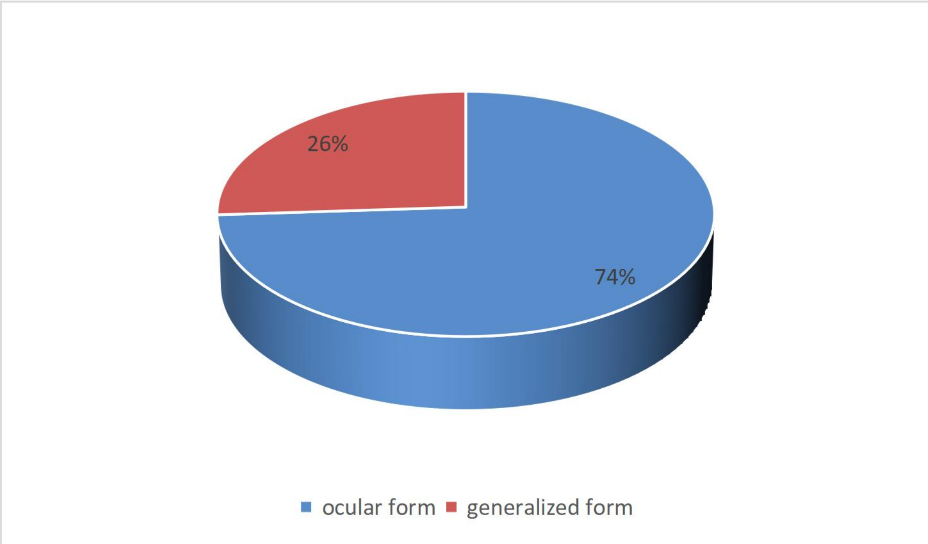

Foto 1. Division of patients according to clinical forms.

AChR-Ab–positive results were identified in approximately 45% of patients, confirming the autoimmune nature of the disease. After treatment with pyridostigmine, clinical improvement was observed in 85% of cases, while thymectomy was performed in 11%. Remission was achieved in 63% of children.

Pediatric myasthenia gravis predominantly presents with an ocular course and is frequently characterized by seronegative variants. Comprehensive diagnostic evaluation is required to ensure accurate diagnosis. Compared with adult forms, pediatric myasthenia gravis generally has a more favorable prognosis. Early pathogenetic therapy significantly improves clinical outcomes and quality of life in affected children.

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