Review

iMedPub Journals
www.imedpub.com

Annals of Clinical and Laboratory Research
ISSN 2386-5180

2025
Vol.13 No.S6:005

Clinical Characteristics and Treatment of Juvenile Myasthenia Gravis

Hideki Shimomura*, Sachi Tokunaga and Naoko Taniguchi

[1]Department of Pediatrics, Hyogo Medical University School of Medicine, Nishinomiya, Hyogo, Japan

***Corresponding author:** Hideki Shimomura, Department of Pediatrics, Hyogo Medical University School of Medicine, Nishinomiya, Hyogo, Japan, E-mail: simo-ped@hyo-med.ac.jp

Received date: 02-Sep-2025, Manuscript No. IPACLR-25-15758; Editor **assigned:** 04-Sep-2025, PreQC No. IPACLR-25-15758 (PQ); **Reviewed:** 17-Sep-2025, QC No IPACLR-25-15758; **Revised:** 23-Sep-2025, Manuscript No. IPACLR-25-15758 (R); **Published:** 02-Oct-2025, DOI: 10.36648/2386-5180.25.13.S6.005

Citation: Shimomura H, Tokunaga S, Taniguchi N (2025) Clinical Characteristics and Treatment of Juvenile Myasthenia Gravis. Ann Clin Lab Vol. 13 No.S6:005

Description

Juvenile Myasthenia Gravis (JMG) is a rare autoimmune neuromuscular disorder characterized by the onset of myasthenia gravis before 18 years of age, with clinical features and management strategies that overlap with and diverge from those of adult-onset diseases. JMG most commonly presents with ocular symptoms, such as ptosis and diplopia, but can progress to generalized muscle weakness, including bulbar and respiratory involvement [1,2]. The prevalence of isolated ocular symptoms is higher in prepubertal children and the rate of Acetylcholine Receptor Antibody (AChR-Ab) positivity is lower than that in adults, with significant ethnic and age-related variability. Spontaneous remission is more frequent in children; however, the risk of generalization and myasthenic crisis remains, especially in children with higher AChR-Ab titers or generalized onset [1-3].

Treatment is typically adapted from adult guidelines, with pyridostigmine as the first-line symptomatic therapy, followed by corticosteroids and other immunosuppressants for more severe or refractory cases [4,5]. Thymectomy is considered in selected patients, particularly those with generalized disease or thymoma and recent studies suggest that minimally invasive approaches are safe and may improve outcomes [6]. Newer immunomodulatory agents, such as rituximab and complement inhibitors (eculizumab, ravulizumab), are emerging as options for refractory cases, although pediatric-specific data remain limited [5,7,8]. Despite the favorable long-term prognosis, especially for ocular JMG, considerable heterogeneity exists in the disease course, response to therapy and risk of comorbid autoimmune conditions.

Epidemiology and Clinical Presentation

JMG is rare, with incidence rates ranging from 1 to 5 per million children per year and prevalence estimates vary by region and ethnicity [1,2,9]. Ocular Myasthenia Gravis (MG) is four times more frequent in Asian children and two to three times more frequent in children of African ancestry than in Europeans. Asian children are more likely to have purely ocular Myasthenia Gravis (MG), while European children often progress to generalized disease [10]. Most cases present before puberty, with a slight female predominance at postpubertal onset. Ocular symptoms (ptosis and diplopia) are the most common initial presentations, especially in prepubertal children and Asian populations, whereas generalized weakness is more frequent in older children and among certain ethnic groups. These symptoms tend to exhibit diurnal variations, with exacerbations during the evening hours. However, this manifestation may be less pronounced in pediatric patients [11]. Symptoms worsen with activity and improve with rest. AChR-Ab positivity is observed in 40–80% of cases, with lower rates in younger children and in some ethnicities. Muscle-Specific Kinase (MuSK) antibodies are rare but associated with more severe disease [1,2,9,12]. In some cases, especially in young children, symptoms may be atypical or overlap with other neuromuscular disorders, making early diagnosis challenging [13].

Diagnosis, Course and Prognostic Factors

The diagnostic tests included serological, electrophysiological and pharmacological tests. Approximately 79% of JMG cases are positive for AChR-Abs; however, positivity rates are lower in prepubertal children. MuSK antibodies may be present in patients who test negative for AChR antibodies [2,13]. Repetitive nerve stimulation and single-fiber electromyography can demonstrate impaired neuromuscular transmission, supporting the diagnosis [13]. Improvement of symptoms after the administration of anticholinesterase agents (neostigmine or pyridostigmine) is a supportive pharmacological test. Spontaneous remission occurs in a minority of patients (3-35%), more commonly in ocular JMG and prepubertal onset, but relapses are possible. Generalization from ocular to generalized MG occurs in 10-25% of cases, typically within the first 2 years. The risk factors for generalization and poor outcomes include higher AChR-Ab titers, generalized onset and longer disease duration before treatment. Comorbid autoimmune diseases, particularly thyroid disorders, are reported in up to one-third of patients [2,3,9,14].

Treatment Modalities and Outcomes

The first-line therapy is pyridostigmine with corticosteroids (prednisone/prednisolone) for insufficient response or generalized disease. Immunosuppressants, such as azathioprine, mycophenolate mofetil and tacrolimus, are used as steroid-sparing agents or in refractory cases [1-4,15]. Although oral medications are often effective, the available evidence regarding dosage escalation methods, treatment duration and discontinuation protocols remains limited. Our previous study indicated that at least 33 months of oral treatment is required [16]. Rituximab has shown efficacy and good tolerability, particularly when used early in antibody-positive generalized JMG [5,17]. Eculizumab and ravulizumab complement inhibitors have demonstrated benefits in small pediatric cohorts with refractory disease [7,8]. Thymectomy is considered for generalized JMG, thymoma, or persistent symptoms, with evidence supporting improved remission rates and steroid-sparing effects, especially with early intervention and minimally invasive approaches [4,6,18]. Plasma exchange and intravenous immunoglobulin are reserved for myasthenic crises or perioperative management, with plasmapheresis showing a more consistent response [2,4,12].

Long-Term Outcomes and Complications

The long-term prognosis is favorable, with 24-66% of patients achieving complete or pharmacologic remission and most patients experience significant improvement. Ocular JMG has a higher remission rate and a lower risk of generalization than generalized JMG [1,2,9,19]. Adverse effects of therapy, particularly growth retardation due to steroids and the risks of immuno- suppression, are important considerations in children [2-4]. Persistent ophthalmoplegia and amblyopia are notable complications, especially in African and Asian populations [1,14,20].

Conclusion

JMG is a rare but treatable autoimmune disorder with distinct clinical features and favorable outcomes, particularly in ocular presentation. Treatment was adapted from adult protocols, with pyridostigmine, corticosteroids, immunosuppressants and thymectomy as mainstays and newer biologics have emerged for refractory cases. However, the evidence base is limited by the rarity of the disease and the lack of pediatric-specific trials, underscoring the need for collaborative research and standardized guidelines for its management. Despite these advances, significant gaps remain in our understanding of the optimal timing and choice of immunosuppressive agents, the long-term safety of newer biologics and the best strategies for preventing and managing complications, such as persistent ophthalmoplegia and steroid side effects. A need exists for more robust data on ethnic and age-related differences in the course of disease and response to therapy.

References

1. Huang X, Li Y, Feng H, Chen P, Liu W (2018) Clinical Characteristics of Juvenile Myasthenia Gravis in Southern China. Front Neurol 9:77.
2. Lin Y, Kuang Q, Li H, Liang B, Lu J, et al. (2023) Outcome and clinical features in juvenile myasthenia gravis: A systematic review and meta-analysis. Front Neurol 14:1119294.
3. Yang L, Tang Y, He F, Zhang C, Kessi M, et al. (2022) Clinical characteristics and outcome predictors of a Chinese childhood-onset myasthenia gravis cohort. Front Pediatr 10:996213.
4. O'Connell K, Ramdas S, Palace J (2020) Management of Juvenile Myasthenia Gravis. Front Neurol 11:743.
5. Molimard A, Gitiaux C, Barnerias C, Audic F, Isapof A, et al. (2022) Rituximab Therapy in the Treatment of Juvenile Myasthenia Gravis: The French Experience. Neurology 98:e2368-2376.
6. Ng WC, Hartley L (2021) Effectiveness of thymectomy in juvenile myasthenia gravis and clinical characteristics associated with better outcomes. Neuromuscul Disord NMD 31:1113-1123.
7. Ramdas S, Painho T, Vanegas MI, Famili DT, Lim MJ, et al. (2024) Targeted Treatments for Myasthenia Gravis in Children and Adolescents. Paediatr Drugs 26:719-740.
8. Brandsema JF, Ginsberg M, Hoshino H, Mimaki M, Nagata S, et al. (2024) Eculizumab in Adolescent Patients With Refractory Generalized Myasthenia Gravis: A Phase 3, Open-Label, Multicenter Study. Pediatr Neurol 156:198-207.
9. Lee HN, Kang H-C, Lee JS, Kim HD, Shin HY, et al. (2016) Juvenile Myasthenia Gravis in Korea: Subgroup Analysis According to Sex and Onset Age. J Child Neurol 31:1561-1568.
10. Heckmann JM, Europa TA, Soni AJ, Nel M (2022) The Epidemiology and Phenotypes of Ocular Manifestations in Childhood and Juvenile Myasthenia Gravis: A Review. Front Neurol 13:834212.
11. Dresser L, Wlodarski R, Rezania K, Soliven B (2021) Myasthenia Gravis: Epidemiology, Pathophysiology and Clinical Manifestations. J Clin Med 10:2235.
12. VanderPluym J, Vajsar J, Jacob FD, Mah JK, Grenier D, et al. (2013) Clinical characteristics of pediatric myasthenia: A surveillance study. Pediatrics 132:e939-944.
13. Della Marina A, Trippe H, Lutz S, Schara U (2014) Juvenile myasthenia gravis: Recommendations for diagnostic approaches and treatment. Neuropediatrics 45:75-83.
14. Vanikieti K, Lowwongngam K, Padungkiatsagul T, Visudtibhan A, Poonyathalang A (2018) Juvenile Ocular Myasthenia Gravis: Presentation and Outcome of a Large Cohort. Pediatr Neurol 87:36-41.
15. Wang G, Kessi M, Huang X, Zhang W, Zhang C, et al. (2024) Treatment of juvenile myasthenia gravis with tacrolimus: A cohort study. Eur J Neurol 31:e16466.
16. Maeda M, Shimomura H, Tokunaga S, Taniguchi N, Lee T, et al. (2024) Clinical Characteristics and Treatment of Juvenile Myasthenia Gravis-A Single-Center Experience. Child Basel Switz 11:572.
17. Zingariello CD, Elder ME, Kang PB (2020) Rituximab as Adjunct Maintenance Therapy for Refractory Juvenile Myasthenia Gravis. Pediatr Neurol 111:40-43.

18. Kim AG, Upah SA, Brandsema JF, Yum SW, Blinman TA (2019) Thoracoscopic thymectomy for juvenile myasthenia gravis. Pediatr Surg Int 35:603-610.

19. Popperud TH, Boldingh MI, Rasmussen M, Kerty E (2017) Juvenile myasthenia gravis in Norway: Clinical characteristics, treatment, and long-term outcome in a nationwide population-based cohort. Eur J Paediatr Neurol 21:707-714.

20. Ortiz S, Borchert M (2008) Long-term outcomes of pediatric ocular myasthenia gravis. Ophthalmology 115:1245-1248.e1.