

LRP4 Autoimmune Reactivity in a Diverse Double-Seronegative Myasthenia Gravis Patient Population

Background

- Myasthenia gravis (MG) is an autoimmune disease that causes defective transmission of nerve impulses to muscle cells.
- Many MG patients test positive for autoantibodies to the acetylcholine receptor (AChR) or to muscle-specific kinase (MuSK); MG patients who have neither are considered double seronegative.¹
- Double-seronegative MG patients who have autoantibodies to the low-density lipoprotein receptor-related protein 4 (LRP4) may have a milder prognosis and better response to treatment than other MG patients.
- Autoantibodies to LRP4 have been reported at rates ranging from 0% to 54% among double-seronegative MG patients; the rate may depend in part on patient demographics.
- **Objective:** The investigators of this study examined the frequency of LRP4 autoantibodies in a demographically diverse population of double-seronegative MG patients in the United States.

Methods

- Serum samples were analyzed for LRP4 autoimmune reactivity with a dual-labeled immunofluorescence assay.
- Analysis was carried out on samples from
 - 150 double-seronegative MG patients (mean age, 60 y; range, 10-92 y)
 - 101 apparently healthy individuals (mean age, 38 y; range, 19-69 y)

Results

- LRP4 autoantibodies were found in
 - 3% (4 of 150) of samples from double-seronegative MG patients
 - 0% (0 of 101) of samples from apparently healthy individuals
- Of the 4 double-seronegative MG patients positive for LRP4 autoantibodies
 - One was a 58-year-old man with generalized MG.
 - One was a 50-year-old woman with isolated ptosis.
 - Two had no available clinical information.

Conclusions

- In this demographically diverse group of double-seronegative MG patients, the prevalence of LRP4 autoantibodies was in the low end of the previously reported range.
- Additional information on distinguishing clinical features of LRP4 antibody-positive individuals with MG will be needed to assess implications for prognosis and treatment response.

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Webpage

<http://submissions.mirasmart.com/Verify/AAN2017/submission/template/radA8953.pdf>

References

1. Phillips WD, Vincent A. *F1000Res.* 2016;5. pii: F1000 Faculty Rev-1513.