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Function and structure of the eye muscles in myasthenia gravis: novel methods to aid in diagnosis and understanding of pathophysiology

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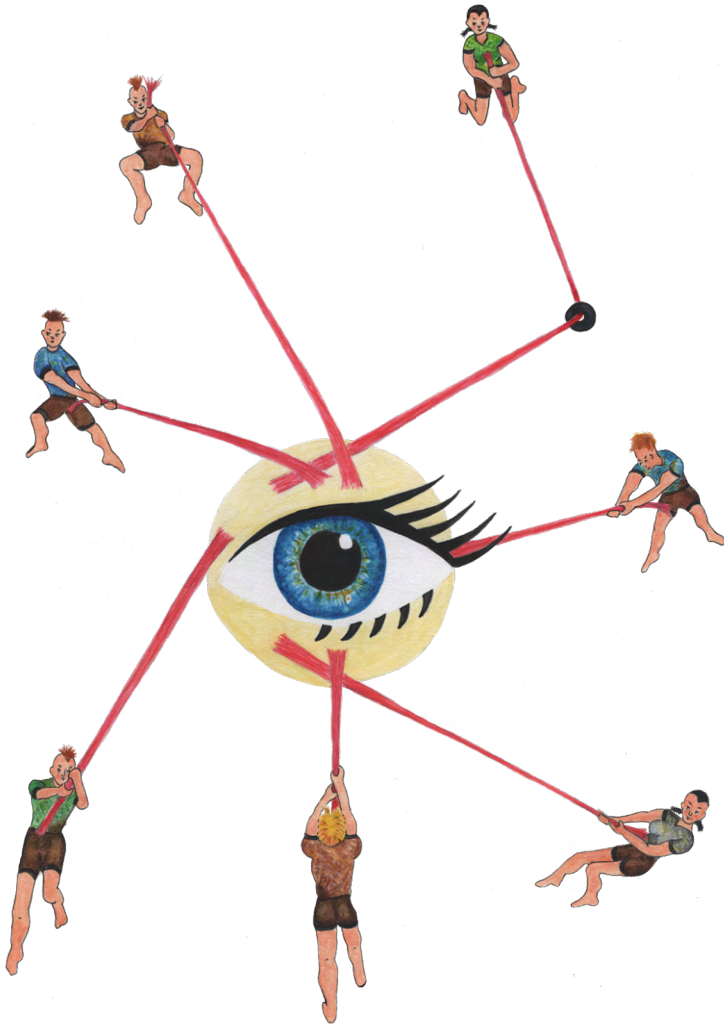
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References

1. Grob D, Brunner N, Namba T, Pagala M. Lifetime course of myasthenia gravis. *Muscle Nerve*. 2008;37(2):141-149. doi:10.1002/mus.20950
2. Gilhus NE, Tzartos S, Evoli A, Palace J, Burns TM, Verschuuren JJGM. Myasthenia gravis. *Nat Rev Dis Primers*. 2019;5(1):30. doi:10.1038/s41572-019-0079-y
3. Fortin E, Cestari DM, Weinberg DH. Ocular myasthenia gravis: An update on diagnosis and treatment. *Curr Opin Ophthalmol*. 2018;29(6):477-484. doi:10.1097/ICU.0000000000000526
4. Punga AR, Maddison P, Heckmann JM, Guptill JT, Evoli A. Epidemiology, diagnostics, and biomarkers of autoimmune neuromuscular junction disorders. *Lancet Neurol*. 2022;21(2):176-188. doi:10.1016/S1474-4422(21)00297-0
5. Gilhus NE, Verschuuren JJ. Myasthenia gravis: Subgroup classification and therapeutic strategies. *Lancet Neurol*. 2015;14(10):1023-1036. doi:10.1016/S1474-4422(15)00145-3
6. Huijbers MG, Marx A, Plomp JJ, Le Panse R, Phillips WD. Advances in the understanding of disease mechanisms of autoimmune neuromuscular junction disorders. *Lancet Neurol*. 2022;21(2):163-175. doi:10.1016/S1474-4422(21)00357-4
7. Verschuuren JJ, Palace J, Murai H, Tannemaat MR, Kaminski HJ, Bril V. Advances and ongoing research in the treatment of autoimmune neuromuscular junction disorders. *Lancet Neurol*. 2022;21(2):189-202. doi:10.1016/S1474-4422(21)00463-4/ATTACHMENT/30BA1066-C5F6-47B0-9DF6-0F4E2013D4C4/MMC1.PDF
8. Mantegazza R, Antozzi C. When myasthenia gravis is deemed refractory: Clinical signposts and treatment strategies. *Ther Adv Neurol Disord*. 2018;11. doi:10.1177/1756285617749134
9. Heckmann JM, Nel M. A unique subphenotype of myasthenia gravis. *Ann N Y Acad Sci*. 2018;1412(1):14-20. doi:10.1111/nyas.13471
10. Benatar M. A systematic review of diagnostic studies in myasthenia gravis. *Neuromuscular Disorders*. 2006;16(7):459-467. doi:10.1016/j.nmd.2006.05.006
11. Smith S V., Lee AG. Update on Ocular Myasthenia Gravis. *Neurol Clin*. 2017;35(1):115-123. doi:10.1016/j.ncl.2016.08.008
12. Verschuuren JJGM, Palace J, Erik Gilhus N. Clinical aspects of myasthenia explained. *Autoimmunity*. 2010;43(5-6):344-352. doi:10.3109/08916931003602130
13. Gratton SM, Herro A, Bermudez-Magner JA, Guy J. Atrophy and Fibrosis of Extra-Ocular Muscles in Anti-Acetylcholine Receptor Antibody Myasthenia Gravis. *Open J Ophthalmol*. 2014;04(04):117-119. doi:10.4236/ojoph.2014.44019
14. Ortube MC, Bholra R, Demer JL. Orbital Magnetic Resonance Imaging of Extraocular Muscles in Chronic Progressive External Ophthalmoplegia: Specific Diagnostic Findings. *Journal of AAPOS*. 2006;10(5):414-418. doi:10.1016/j.jaapos.2006.04.012
15. Pedapati R, Ravishankar U, Hazeena P, Shanmugam S, Chandrasekharan A, Venkatasubramanian S. Extraocular muscle atrophy in myasthenia gravis. *Ann Indian Acad Neurol*. 2022;25(3):483. doi:10.4103/AIAN.AIAN_1039_21
16. Pedapati R, Ravishankar U, Hazeena P, Shanmugam S, Chandrasekharan A, Venkatasubramanian S. Extraocular Muscle Atrophy in Myasthenia Gravis. *Ann Indian Acad Neurol*. 2022;0(0):0. doi:10.4103/aian.aian_1039_21
17. Velonakis G, Papadopoulos VE, Karavasilis E, Filippiadis DK, Zouvelou V. MRI evidence of extraocular muscle atrophy and fatty replacement in myasthenia gravis. *Neuroradiology*. 2021;63(9):1531-1538. doi:10.1007/S00234-021-02753-4/TABLES/3

18. Farrugia ME, Robson MD, Clover L, et al. MRI and clinical studies of facial and bulbar muscle involvement in MuSK antibody-associated myasthenia gravis. *Brain*. 2006;129(6):1481-1492. doi:10.1093/brain/awl095
19. Nikolić A V., Bačić GG, Daković M, et al. Myopathy, muscle atrophy and tongue lipid composition in MuSK myasthenia gravis. *Acta Neurol Belg*. 2015;115(3):361-365. doi:10.1007/s13760-014-0364-1
20. Oosterhuis H, Bethlem J. Neurogenic muscle involvement in myasthenia gravis. A clinical and histopathological study. *J Neurol Neurosurg Psychiatry*. 1973;36(2):244-254. doi:10.1136/jnnp.36.2.244
21. Vilquin JT, Bayer AC, Le Panse R, Berrih-Aknin S. The Muscle Is Not a Passive Target in Myasthenia Gravis. *Front Neurol*. 2019;10. doi:10.3389/fneur.2019.01343
22. Wolfe GI, Herbelin L, Nations SP, Foster B, Bryan WW, Barohn RJ. Myasthenia gravis activities of daily living profile. *Neurology*. 1999;52(7):1487-1489. doi:10.1212/wnl.52.7.1487
23. Jaretzki A, Barohn RJ, Ernstoff RM, et al. Myasthenia gravis: Recommendations for clinical research standards. *Neurology*. 2000;55(1):16-23. doi:10.1212/WNL.55.1.16
24. Howard JF, Freimer M, O'Brien F, Wang JJ, Collins SR, Kissel JT. QMG and MG-ADL correlations: Study of eculizumab treatment of myasthenia gravis. *Muscle Nerve*. 2017;56(2):328-330. doi:10.1002/MUS.25529
25. Howard JF, Utsugisawa K, Benatar M, et al. Safety and efficacy of eculizumab in anti-acetylcholine receptor antibody-positive refractory generalised myasthenia gravis (REGAIN): a phase 3, randomised, double-blind, placebo-controlled, multicentre study. *Lancet Neurol*. 2017;16(12):976-986. doi:10.1016/S1474-4422(17)30369-1
26. Howard JF, Bril V, Vu T, et al. Safety, efficacy, and tolerability of efgartigimod in patients with generalised myasthenia gravis (ADAPT): a multicentre, randomised, placebo-controlled, phase 3 trial. *Lancet Neurol*. 2021;20(7):526-536. doi:10.1016/S1474-4422(21)00159-9
27. Wong SH, Eggenberger E, Cornblath W, et al. Preliminary Findings of a Dedicated Ocular Myasthenia Gravis Rating Scale: The OMGRate. *Neuro-Ophthalmology*. 2020;44(3):148-156. doi:10.1080/01658107.2019.1660686
28. Oh SJ, Eslami N, Nishihira T, et al. Electrophysiological and clinical correlation in myasthenia gravis. *Ann Neurol*. 1982;12(4):348-354. doi:10.1002/ana.410120406
29. Piker EG, Jacobson GP, Burkard RF, McCaslin DL, Hood LJ. Effects of age on the tuning of the cVEMP and oVEMP. *Ear Hear*. 2013;34(6). doi:10.1097/AUD.0b013e31828fc9f2
30. Weber KP, Rosengren SM, Michels R, Sturm V, Straumann D, Landau K. Single motor unit activity in human extraocular muscles during the vestibulo-ocular reflex. *Journal of Physiology*. 2012;590(13):3091-3101. doi:10.1113/jphysiol.2011.226225
31. Valko Y, Rosengren SM, Jung HH, Straumann D, Landau K, Weber KP. Ocular vestibular evoked myogenic potentials as a test for myasthenia gravis. *Neurology*. 2016;86(7):660-668. doi:10.1212/WNL.0000000000002383
32. de Meel RHP, Keene KR, Wirth MA, et al. Repetitive ocular vestibular evoked myogenic potentials in myasthenia gravis. *Neurology*. 2020;94(16):E1693-E1701. doi:10.1212/WNL.0000000000009306
33. UK HS. SYNOPTOPHORE Information guide. https://www.haag-streit.com/fileadmin/Haag-Streit_UK/Downloads/CCO_downloads/Synoptophore_downloads/Synoptophore_Info_Guide.pdf

34. Guo J, Li X, Ma R, Qian J. Correlation between uniocular deviation and duction changes following different decompression surgeries in thyroid eye disease. *BMC Ophthalmol.* 2021;21(1). doi:10.1186/s12886-021-01892-9
35. Roper-Hall G. The Hess Screen Test. *American Orthoptic Journal.* 2006;56(1):166-174. doi:10.3368/aoj.56.1.166
36. Strijkers GJ, Araujo ECA, Azzabou N, et al. Exploration of new contrasts, targets, and MR imaging and spectroscopy techniques for neuromuscular disease-A workshop report of working group 3 of the biomedicine and molecular biosciences COST action BM1304 MYO-MRI. *J Neuromuscul Dis.* 2019;6(1):1-30. doi:10.3233/JND-180333
37. Burakiewicz J, Sinclair CDJ, Fischer D, Walter GA, Kan HE, Hollingsworth KG. Quantifying fat replacement of muscle by quantitative MRI in muscular dystrophy. *J Neurol.* 2017;264(10):2053-2067. doi:10.1007/s00415-017-8547-3
38. Naarding KJ, Reyngoudt H, Van Zwet EW, et al. MRI vastus lateralis fat fraction predicts loss of ambulation in Duchenne muscular dystrophy. *Neurology.* 2020;94(13):E1386-E1394. doi:10.1212/WNL.0000000000008939
39. Carlier PG, Marty B, Scheidegger O, et al. Skeletal Muscle Quantitative Nuclear Magnetic Resonance Imaging and Spectroscopy as an Outcome Measure for Clinical Trials. *J Neuromuscul Dis.* 2016;3(1):1-28. doi:10.3233/JND-160145
40. Dixon WT. Simple proton spectroscopic imaging. *Radiology.* 1984;153(1):189-194. doi:10.1148/radiology.153.1.6089263
41. Marty B, Baudin PY, Reyngoudt H, et al. Simultaneous muscle water T2 and fat fraction mapping using transverse relaxometry with stimulated echo compensation. *NMR Biomed.* 2016;29(4):431-443. doi:10.1002/nbm.3459
42. Tardif-de Géry S, Vilquin JT, Carlier P, et al. Muscular transverse relaxation time measurement by magnetic resonance imaging at 4 Tesla in normal and dystrophic dy/dy and $dy(2j)/dy(2j)$ mice. *Neuromuscul Disord.* 2000;10(7):507-513. doi:10.1016/S0960-8966(00)00122-X
43. Hardy PA, Henkelman RM, Bishop JE, Poon ECS, Plewes DB. Why fat is bright in rare and fast spin-echo imaging. *Journal of Magnetic Resonance Imaging.* 1992;2(5):533-540. doi:10.1002/jmri.1880020511
44. Niendorf T, Beenakker JWM, Langner S, et al. Ophthalmic Magnetic Resonance Imaging: Where Are We (Heading To)? *Curr Eye Res.* 2021;46(9):1251-1270. doi:10.1080/0/02713683.2021.1874021
45. Beenakker JWM, van Rijn GA, Luyten GPM, Webb AG. High-resolution MRI of uveal melanoma using a microcoil phased array at 7 T. *NMR Biomed.* 2013;26(12):1864-1869. doi:10.1002/nbm.3041
46. Lindner T, Langner S, Graessl A, et al. High spatial resolution in vivo magnetic resonance imaging of the human eye, orbit, nervus opticus and optic nerve sheath at 7.0 Tesla. *Exp Eye Res.* 2014;125:89-94. doi:10.1016/j.exer.2014.05.017
47. Heidary G, Engle EC, Hunter DG. Congenital fibrosis of the extraocular muscles. *Semin Ophthalmol.* 2008;23(1):3-8. doi:10.1080/08820530701745181
48. Al Othman B, Raabe J, Kini A, Lee AG. Update: The Miller Fisher variants of Guillain-Barré syndrome. *Curr Opin Ophthalmol.* 2019;30(6):462-466. doi:10.1097/ICU.0000000000000611

49. Filho ARG, Faccenda PG, Estacia CT, Correa BS, Curi I. Tolosa-Hunt Syndrome. *Rev Bras Oftalmol.* 2018;77(5):289-291. doi:10.5935/0034-7280.20180063
50. Mansukhani SA, Bothun ED, Diehl NN, Mohney BG. Incidence and Ocular Features of Pediatric Myasthenias. *Am J Ophthalmol.* 2019;200:242-249. doi:10.1016/j.ajo.2019.01.004
51. Parmar H, Ibrahim M. Extrathyroidal Manifestations of Thyroid Disease: Thyroid Ophthalmopathy. *Neuroimaging Clin N Am.* 2008;18(3):527-536. doi:10.1016/j.nic.2008.03.003
52. McClelland C, Manousakis G, Lee MS. Progressive External Ophthalmoplegia. *Curr Neurol Neurosci Rep.* 2016;16(6):1-10. doi:10.1007/s11910-016-0652-7
53. Renard D, Ferraro A, Lorenzini MC, et al. Orthoptic and video-oculographic analyses in oculopharyngeal muscular dystrophy. *Muscle Nerve.* 2015;52(4):554-558. doi:10.1002/mus.24600
54. Díaz-Manera J, Luna S, Roig C. Ocular ptosis: Differential diagnosis and treatment. *Curr Opin Neurol.* 2018;31(5):618-627. doi:10.1097/WCO.0000000000000600
55. Holmes JM, Leske DA, Kupersmith MJ. New methods for quantifying diplopia. *Ophthalmology.* 2005;112(11):2035-2039. doi:10.1016/j.ophtha.2005.06.013
56. Latting MW, Huggins AB, Marx DP, Giacometti JN. Clinical Evaluation of Blepharoptosis: Distinguishing Age-Related Ptosis from Masquerade Conditions. *Semin Plast Surg.* 2017;31(1):5-16. doi:10.1055/s-0037-1598188
57. Ozgen A, Ariyurek M. Normative measurements of orbital structures using CT. *American Journal of Roentgenology.* 1998;170(4):1093-1096. doi:10.2214/ajr.170.4.9530066
58. Park NR, Moon JH, Lee JK. Hertel exophthalmometer versus computed tomography scan in proptosis estimation in thyroid-associated orbitopathy. *Clinical Ophthalmology.* 2019;13:1461-1467. doi:10.2147/OPTH.S216838
59. Bau V, Zierz S. Update on chronic progressive external ophthalmoplegia. *Strabismus.* 2005;13(3):133-142. doi:10.1080/09273970500216432
60. Engle EC, Goumnerov BC, McKeown CA, et al. Oculomotor nerve and muscle abnormalities in congenital fibrosis of the extraocular muscles. *Ann Neurol.* 1997;41(3):314-325. doi:10.1002/ana.410410306
61. Bosley TM, Oystreck DT, Robertson RL, Al Awad A, Abu-Amero K, Engle EC. Neurological features of congenital fibrosis of the extraocular muscles type 2 with mutations in PHOX2A. *Brain.* 2006;129(9):2363-2374. doi:10.1093/brain/awl161
62. Marengo M, Macchi I, Macchi I, Galassi E, Massaro-Giordano M, Lambiase A. Clinical presentation and management of congenital ptosis. *Clinical Ophthalmology.* 2017;11:453-463. doi:10.2147/OPTH.S111118
63. Watanabe K, Hagura R, Akanuma Y, et al. Characteristics of cranial nerve palsies in diabetic patients. *Diabetes Res Clin Pract.* 1990;10(1):19-27. doi:10.1016/0168-8227(90)90077-7
64. Lacey B, Chang W, Rootman J. Nonthyroid causes of extraocular muscle disease. *Surv Ophthalmol.* 1999;44(3):187-213. doi:10.1016/S0039-6257(99)00101-0
65. Nagy E V., Toth J, Kaldi I, et al. Graves' ophthalmopathy: Eye muscle involvement in patients with diplopia. *Eur J Endocrinol.* 2000;142(6):591-597. doi:10.1530/eje.0.1420591

66. Fraser CL, Skalicky SE, Gurbaxani A, McCluskey P. Ocular myositis. *Curr Allergy Asthma Rep.* 2013;13(3):315-321. doi:10.1007/s11882-012-0319-7
67. Goto H, Takahira M, Azumi A. Diagnostic criteria for IgG4-related ophthalmic disease. *Jpn J Ophthalmol.* 2015;59(1). doi:10.1007/s10384-014-0352-2
68. Montagnese F, Wenninger S, Schoser B. "Orbiting around" the orbital myositis: clinical features, differential diagnosis and therapy. *J Neurol.* 2016;263(4):631-640. doi:10.1007/s00415-015-7926-x
69. Gladstone JP, Dodick DW. Painful ophthalmoplegia: Overview with a focus on Tolosa-Hunt syndrome. *Curr Pain Headache Rep.* 2004;8(4):321-329. doi:10.1007/s11916-004-0016-x
70. Nishikawa N, Kawaguchi Y, Konno A, Kitani Y, Takei H, Yanagi Y. Primary isolated amyloidosis in the extraocular muscle as a rare cause of ophthalmoplegia: A case report and literature review. *Am J Ophthalmol Case Rep.* 2021;22:101052. doi:10.1016/j.ajoc.2021.101052
71. Jenkins PO, Soper C, Mackinnon AD, O'Sullivan E, Nitkunan A. Systemic lupus erythematosus presenting as orbital myositis. *Neuro-Ophthalmology.* 2014;38(5):264-267. doi:10.3109/01658107.2014.923915
72. Chan AJ, Rai AS, Lake S. Orbital myositis in systemic lupus erythematosus: A case report and literature review. *Eur J Rheumatol.* 2020;7(3):135-137. doi:10.5152/eurjrheum.2020.19217
73. Kim JS, Scawn RL, Lee BW, Lin JH, Korn BS, Kikkawa DO. Masquerading Orbital Sarcoidosis with Isolated Extraocular Muscle Involvement. *Open Ophthalmol J.* 2016;10(1):140. doi:10.2174/1874364101610010140
74. Pimentel R, Lago P, Pedroto I. Recurrent orbital myositis as an extra-intestinal manifestation of Crohn's disease. *J Crohns Colitis.* 2012;6(9):958-959. doi:10.1016/j.crohns.2012.05.018
75. Ramalho J, Castillo M. Imaging of orbital myositis in Crohn's disease. *Clin Imaging.* 2008;32(3):227-229. doi:10.1016/j.clinimag.2007.09.012
76. Bourikas LA, Roussomoustakaki M, Papadaki E, et al. A case of orbital myositis preceding the intestinal symptoms of Crohn's disease. *J Crohns Colitis.* 2010;4(3):349-350. doi:10.1016/j.crohns.2010.05.005
77. McNab AA. Orbital myositis: A comprehensive review and reclassification. *Ophthalmic Plast Reconstr Surg.* Published online 2020:109-117. doi:10.1097/IOP.0000000000001429
78. Nair AG, Patil-Chhablani P, Venkatramani D V., Gandhi RA. Ocular myasthenia gravis: A review. *Indian J Ophthalmol.* 2014;62(10):985-991. doi:10.4103/0301-4738.145987
79. Jeannet PY, Bassez G, Eymard B, et al. *Clinical and Histologic Findings in Autosomal Centronuclear Myopathy.* Vol 62.; 2004. doi:10.1212/01.WNL.0000124388.67003.56
80. Thornton CA. Myotonic dystrophy. *Neurol Clin.* 2014;32(3):705-719. doi:10.1016/j.ncl.2014.04.011
81. Choi SH, Yang HK, Hwang JM, Park KS. Ocular Findings of Myotonic Dystrophy Type 1 in the Korean Population. *Graefe's Archive for Clinical and Experimental Ophthalmology.* 2016;254(6):1189-1193. doi:10.1007/s00417-016-3266-5
82. Thiriez C, Vignal C, Papeix C, Yaici S, Vidailhet M, Roze E. Ophthalmoplegia as the presenting muscle-related manifestation of myotonic dystrophy. *Rev Neurol (Paris).* 2010;166(5):538-541. doi:10.1016/j.neurol.2009.12.004

83. Van Der Beek NAME, De Vries JM, Hagemans MLC, et al. Clinical features and predictors for disease natural progression in adults with Pompe disease: A nationwide prospective observational study. *Orphanet J Rare Dis.* 2012;7(1):88. doi:10.1186/1750-1172-7-88
84. Fischer MD, Budak MT, Bakay M, et al. Definition of the unique human extraocular muscle allotype by expression profiling. *Physiol Genomics.* 2005;22:283-291. doi:10.1152/physiolgenomics.00158.2004
85. Burke G, Cossins J, Maxwell S, et al. Rapsyn mutations in hereditary myasthenia: Distinct early- and late-onset phenotypes. *Neurology.* 2003;61(6):826-828. doi:10.1212/01.WNL.0000085865.55513.AE
86. Finsterer J. Congenital myasthenic syndromes. *Orphanet J Rare Dis.* 2019;14(1):1-22. doi:10.1186/s13023-019-1025-5
87. Ferreira TA, Saraiva P, Genders SW, Buchem M v., Luyten GPM, Beenakker JW. CT and MR imaging of orbital inflammation. *Neuroradiology.* 2018;60(12):1253-1266. doi:10.1007/s00234-018-2103-4
88. Lakerveld M, van der Gijp A. Orbital Muscle Enlargement: What if It's Not Graves' Disease? *Curr Radiol Rep.* 2022;10(2):9-19. doi:10.1007/s40134-022-00392-y
89. Tiegs-Heiden CA, Eckel LJ, Hunt CH, et al. Immunoglobulin G4-related disease of the orbit: Imaging features in 27 patients. *American Journal of Neuroradiology.* 2014;35(7):1393-1397. doi:10.3174/ajnr.A3865
90. Garau LM, Guerrieri D, De Cristofaro F, Bruscolini A, Panzironi G. Extraocular muscle sampled volume in Graves' orbitopathy using 3-T fast spin-echo MRI with iterative decomposition of water and fat sequences. *Acta Radiol Open.* 2018;7(6):205846011878089. doi:10.1177/2058460118780892
91. Lennerstrand G, Tian S, Isberg B, et al. Magnetic resonance imaging and ultrasound measurements of extraocular muscles in thyroid-associated ophthalmopathy at different stages of the disease. *Acta Ophthalmol Scand.* 2007;85(2):192-201. doi:10.1111/j.1600-0420.2006.00807.x
92. Culver EL, Salmon JF, Frith P, Travis SPL. Recurrent posterior scleritis and orbital myositis as extra-intestinal manifestations of Crohn's disease: Case report and systematic literature review. *J Crohns Colitis.* 2008;2(4):337-342. doi:10.1016/j.crohns.2008.06.002
93. Pitceathly RDS, Morrow JM, Sinclair CDJ, et al. Extra-ocular muscle MRI in genetically-defined mitochondrial disease. *Eur Radiol.* 2016;26(1):130-137. doi:10.1007/s00330-015-3801-5
94. Lessell S, Coppeto J, Samet S. Ophthalmoplegia in myotonic dystrophy. *Am J Ophthalmol.* 1971;71(6):1231-1235. doi:10.1016/0002-9394(71)90968-8
95. Demer JL, Clark RA, Engle EC. Magnetic resonance imaging evidence for widespread orbital dysinnervation in congenital fibrosis of extraocular muscles due to mutations in KIF21A. *Invest Ophthalmol Vis Sci.* 2005;46(2):530-539. doi:10.1167/iovs.04-1125
96. Razek AAKA, Maher H, Kasem MA, Helmy E. Imaging of congenital cranial dysinnervation disorders: What radiologist wants to know? *Clin Imaging.* 2021;71:106-116. doi:10.1016/j.clinimag.2020.10.055
97. Chan JW, Orrison WW. Ocular myasthenia: A rare presentation with MuSK antibody and bilateral extraocular muscle atrophy [6]. *British Journal of Ophthalmology.* 2007;91(6):842-843. doi:10.1136/bjo.2006.108498

98. Ricciardi D, Todisco V, Tedeschi G, Cirillo G. Anti-MuSK ocular myasthenia with extrinsic ocular muscle atrophy: a new clinical phenotype? *Neurological Sciences*. 2020;41(1):221-223. doi:10.1007/s10072-019-04038-4
99. Keene KR, van Vught L, van de Velde NM, et al. The feasibility of quantitative MRI of extra-ocular muscles in myasthenia gravis and Graves' orbitopathy. *NMR Biomed*. 2021;34(1):e4407. doi:10.1002/nbm.4407
100. Pakdaman MN. Orbital inflammatory disease: Pictorial review and differential diagnosis. *World J Radiol*. 2014;6(4):106. doi:10.4329/wjr.v6.i4.106
101. Ding ZX, Lip G, Chong V. Idiopathic orbital pseudotumour. *Clin Radiol*. 2011;66(9):886-892. doi:10.1016/j.crad.2011.03.018
102. Tortora F, Cirillo M, Ferrara M, et al. *Disease Activity in Graves' Ophthalmopathy: Diagnosis with Orbital MR Imaging and Correlation with Clinical Score*. Vol 26.; 2013. doi:10.1177/197140091302600509
103. Mombaerts I, Rose GE, Garrity JA. Orbital inflammation: Biopsy first. *Surv Ophthalmol*. 2016;61(5):664-669. doi:10.1016/j.survophthal.2016.03.002
104. Lueangaram S, Tritanon O, Siriyotha S, et al. Radiological characteristics of extraocular muscles in myasthenia gravis patients with ocular manifestations: A case-control study. *Clinical Ophthalmology*. 2021;15:2279-2285. doi:10.2147/OPTH.S280508
105. Kirsch E, Hammer B, von Arx G. Graves' orbitopathy: Current imaging procedures. *Swiss Med Wkly*. 2009;139(43-44):618-623. doi:10.4414/smw.2009.12741
106. Dik WA, Virakul S, van Steensel L. Current perspectives on the role of orbital fibroblasts in the pathogenesis of Graves' ophthalmopathy. *Exp Eye Res*. 2016;142:83-91. doi:10.1016/j.exer.2015.02.007
107. Potgieser PW, de Win MMML, Wiersinga WM, Mourits MP. Natural Course of Mild Graves Orbitopathy: Increase of Orbital Fat But Decrease of Muscle Volume With Increased Muscle Fatty Degeneration During a 4-Year Follow-Up. *Ophthalmic Plast Reconstr Surg*. 2019;35(5):456-460. doi:10.1097/IOP.0000000000001319
108. Sogabe Y, Ohshima KI, Azumi A, et al. Location and frequency of lesions in patients with IgG4-related ophthalmic diseases. *Graefe's Archive for Clinical and Experimental Ophthalmology*. 2014;252(3):531-538. doi:10.1007/s00417-013-2548-4
109. Fischer D, Herasse M, Bitoun M, et al. Characterization of the muscle involvement in dynamin 2-related centronuclear myopathy. *Brain*. 2006;129(6):1463-1469. doi:10.1093/brain/awl071
110. De Meel RHP, Raadsheer WF, Van Zwet EW, Tannemaat MR, Verschuuren JJGM. Ocular Weakness in Myasthenia Gravis: Changes in Affected Muscles are a Distinct Clinical Feature. *J Neuromuscul Dis*. 2019;6(3):369-376. doi:10.3233/JND-190407
111. Almog Y, Ben-David M, Nemet AY. Inferior oblique muscle paresis as a sign of myasthenia gravis. In: *Journal of Clinical Neuroscience*. Vol 25. J Clin Neurosci; 2016:50-53. doi:10.1016/j.jocn.2015.08.026
112. Cleary M, Williams GJ, Metcalfe RA. The pattern of extra-ocular muscle involvement in ocular myasthenia. *Strabismus*. 2008;16(1):11-18. doi:10.1080/15569520701830992
113. Young JD, Leavitt JA. Lambert-Eaton myasthenic syndrome: Ocular signs and symptoms. *Journal of Neuro-Ophthalmology*. 2016;36(1):20-22. doi:10.1097/WNO.0000000000000258

114. Ryu WY, Kim YH, Yoon BA, Park HT, Bae JS, Kim JK. Pattern of extraocular muscle involvements in miller fisher syndrome. *Journal of Clinical Neurology (Korea)*. 2019;15(3):308-312. doi:10.3988/jcn.2019.15.3.308
115. Isen DR, Kline LB. Neuro-ophthalmic manifestations of wernicke encephalopathy. *Eye Brain*. 2020;12:49-60. doi:10.2147/EB.S234078
116. Yellin H. Unique intrafusal and extraocular muscle fibers exhibiting dual actomyosin ATPase activity. *Exp Neurol*. 1969;25(1):153-163. doi:10.1016/0014-4886(69)90078-8
117. Liu JX, Pedrosa Domellöf F. A novel type of multiterminal motor endplate in human extraocular muscles. *Invest Ophthalmol Vis Sci*. 2018;59(1):539-548. doi:10.1167/iovs.17-22554
118. Bishop KN, McClung JR, Goldberg SJ, Shall MS. Anatomic and physiological characteristics of the ferret lateral rectus muscle and abducens nucleus. *J Appl Physiol*. 2007;103(5):1706-1714. doi:10.1152/jappphysiol.00580.2007
119. Sekulic-Jablanovic M, Ullrich ND, Goldblum D, Palmowski-Wolfe A, Zorzato F, Treves S. Functional characterization of orbicularis oculi and extraocular muscles. *Journal of General Physiology*. 2016;147(5):395-406. doi:10.1085/jgp.201511542
120. Sekulic-Jablanovic M, Palmowski-Wolfe A, Zorzato F, Treves S. Characterization of excitation-contraction coupling components in human extraocular muscles. *Biochemical Journal*. 2015;466(1):29-36. doi:10.1042/BJ20140970
121. McLoon LK, Rowe J, Wirtschafter J, McCormick KM. Continuous myofiber remodeling in uninjured extraocular myofibers: Myonuclear turnover and evidence for apoptosis. *Muscle Nerve*. 2004;29(5):707-715. doi:10.1002/mus.20012
122. Kjellgren D, Thornell LE, Andersen J, Pedrosa-Domellöf F. Myosin heavy chain isoforms in human extraocular muscles. *Invest Ophthalmol Vis Sci*. 2003;44(4):1419-1425. doi:10.1167/iovs.02-0638
123. Kitamura K, Cho KH, Jang HS, Murakami G, Yamamoto M, Abe S ichi. Distance between intramuscular nerve and artery in the extraocular muscles: a preliminary immunohistochemical study using elderly human cadavers. *Surgical and Radiologic Anatomy*. 2017;39(1):3-9. doi:10.1007/s00276-016-1642-9
124. Morrow JM, Sinclair CDJ, Fischmann A, et al. MRI biomarker assessment of neuromuscular disease progression: A prospective observational cohort study. *Lancet Neurol*. 2016;15(1):65-77. doi:10.1016/S1474-4422(15)00242-2
125. Europa TA, Nel M, Heckmann JM. A review of the histopathological findings in myasthenia gravis: Clues to the pathogenesis of treatment-resistance in extraocular muscles. *Neuromuscular Disorders*. 2019;29(5):381-387. doi:10.1016/j.nmd.2019.03.009
126. Ferreira TA, Fonk LG, Jaarsma-Coes MG, van Haren GGR, Marinkovic M, Beenakker JWM. MRI of uveal melanoma. *Cancers (Basel)*. 2019;11(3). doi:10.3390/cancers11030377
127. Jaarsma-Coes MG, Marinkovic M, Astreinidou E, et al. Measuring eye deformation between planning and proton beam therapy position using magnetic resonance imaging. *Phys Imaging Radiat Oncol*. 2020;16:33-36. doi:10.1016/j.phro.2020.09.010
128. Chardon JW, Díaz-Manera J, Tasca G, et al. MYO-MRI diagnostic protocols in genetic myopathies. *Neuromuscular Disorders*. 2019;29(11):827-841. doi:10.1016/j.nmd.2019.08.011

129. Howard JF, Vu T, Mantegazza R, et al. MUS MGFA Abstracts: MRI OF THE EXTRA-OCULAR MUSCLES IN MYASTHENIA GRAVIS SHOW SMALL VOLUME AND FAT FRACTION INCREASES. *Muscle Nerve*. 2022;65:S1-S47. doi:10.1002/MUS.27540
130. von Haehling S, Morley JE, Coats AJS, Anker SD. Ethical guidelines for publishing in the journal of cachexia, sarcopenia and muscle: update 2017. *J Cachexia Sarcopenia Muscle*. 2017;8(6):1081-1083. doi:10.1002/jcsm.12261
131. Nadaj-Pakleza A, Richard P, Łusakowska A, et al. Oculopharyngeal muscular dystrophy: Phenotypic and genotypic characteristics of 9 Polish patients. *Neurol Neurochir Pol*. 2009;43(2):113-120.
132. Troost BT, Daroff RB. The ocular motor defects in progressive supranuclear palsy. *Ann Neurol*. 1977;2(5):397-403. doi:10.1002/ana.410020509
133. Straube A, Witt TN. *Oculo-Bulbar Myasthenic Symptoms as the Sole Sign of Tumour Involving or Compressing the Brain Stem*. Vol 237.; 1990. doi:10.1007/BF00315661
134. Liu Y, Wang M, Bian X, et al. Proposed modified diagnostic criteria for recurrent painful ophthalmoplegic neuropathy: Five case reports and literature review. *Cephalalgia*. 2020;40(14):1657-1670. doi:10.1177/0333102420944872
135. Martin TJ. Horner Syndrome: A Clinical Review. *ACS Chem Neurosci*. 2018;9(2):177-186. doi:10.1021/acschemneuro.7b00405
136. Jain R, Sawhney S, Koul RL, Chand P. Tolosa-Hunt syndrome: MRI appearances. *J Med Imaging Radiat Oncol*. 2008;52(5):447-451. doi:10.1111/j.1440-1673.2008.01988.x
137. Schoser B, Eymard B, Datt J, Mantegazza R. Lambert–Eaton myasthenic syndrome (LEMS): a rare autoimmune presynaptic disorder often associated with cancer. *J Neurol*. 2017;264(9):1854-1863. doi:10.1007/s00415-017-8541-9
138. Wirtz PW, Sotodeh M, Nijhuis M, et al. Difference in distribution of muscle weakness between myasthenia gravis and the Lambert-Eaton myasthenic syndrome. *J Neurol Neurosurg Psychiatry*. 2002;73(6):766-768. doi:10.1136/jnnp.73.6.766
139. Luigetti M, Sabatelli M. Cranial botulism. *Neuromuscular Disorders*. 2012;22(11):995-996. doi:10.1016/j.nmd.2012.05.009
140. Chatham-Stephens K, Fleck-Derderian S, Johnson SD, Sobel J, Rao AK, Meaney-Delman D. Clinical Features of Foodborne and Wound Botulism: A Systematic Review of the Literature, 1932-2015. *Clinical Infectious Diseases*. 2017;66(suppl_1):S11-S16. doi:10.1093/cid/cix811
141. Azam L, McIntosh JM. Alpha-conotoxins as pharmacological probes of nicotinic acetylcholine receptors. *Acta Pharmacol Sin*. 2009;30(6):771-783. doi:10.1038/aps.2009.47
142. Olsen TG, Heegaard S. Orbital lymphoma. *Surv Ophthalmol*. 2019;64(1):45-66. doi:10.1016/j.survophthal.2018.08.002
143. Claytor B, Li Y. Challenges in diagnosing coexisting ocular myasthenia gravis and thyroid eye disease. *Muscle Nerve*. 2021;63(5):631-639. doi:10.1002/mus.27118
144. Melcescu E, Horton WB, Kim D, et al. Graves Orbitopathy: Update on Diagnosis and Therapy. *South Med J*. 2014;107(1):34-43. doi:10.1097/SMJ.0000000000000038
145. Marinò M, Ionni I, Lanzolla G, et al. Orbital diseases mimicking graves' orbitopathy: a long-standing challenge in differential diagnosis. *J Endocrinol Invest*. 2020;43(4):401-411. doi:10.1007/s40618-019-01141-3

146. Xian J, Zhang Z, Wang Z, et al. Value of MR imaging in the differentiation of benign and malignant orbital tumors in adults. *Eur Radiol.* 2010;20(7):1692-1702. doi:10.1007/s00330-009-1711-0
147. Fatima Z, Ichikawa T, Ishigame K, et al. Orbital masses: The usefulness of diffusion-weighted imaging in lesion categorization. *Clin Neuroradiol.* 2014;24(2):129-134. doi:10.1007/s00062-013-0234-x
148. El-Sayed Mojahed M, Thabet EM, El-Khateeb MG, Elsayed Morgan A. Ocular vestibular evoked myogenic potential in patients with myasthenia gravis: A prospective clinical study. *Auris Nasus Larynx.* 2018;45(3):407-411. doi:10.1016/j.anl.2017.06.001
149. Decock CE, De Baere EE, Bauters W, et al. Insights into levator muscle dysfunction in a cohort of patients with molecularly confirmed blepharophimosis-ptosis-epicanthus inversus syndrome using high-resolution imaging, anatomic examination, and histopathologic examination. *Archives of Ophthalmology.* 2011;129(12):1564-1569. doi:10.1001/archophthalmol.2011.348
150. Xia S, Li RL, Li YP, Qian XH, Chong V, Qi J. MRI findings in Duane's ocular retraction syndrome. *Clin Radiol.* 2014;69(5). doi:10.1016/j.crad.2013.12.010
151. Thacker NM, Velez FG, Demer JL, Rosenbaum AL. Superior oblique muscle involvement in thyroid ophthalmopathy. *Journal of AAPOS.* 2005;9(2):174-178. doi:10.1016/j.jaapos.2004.12.005
152. Kupersmith MJ, Latkany R, Homel P. Development of generalized disease at 2 years in patients with ocular myasthenia gravis. *Arch Neurol.* 2003;60(2):243-248. doi:10.1001/archneur.60.2.243
153. Padua L, Stalberg E, Lomonaco M, Evoli A, Batocchi A, Tonali P. SFEMG in ocular myasthenia gravis diagnosis. *Clinical Neurophysiology.* 2000;111(7):1203-1207. doi:10.1016/S1388-2457(00)00307-2
154. Venhovens J, Meulstee J, Verhagen WIM. Vestibular evoked myogenic potentials (VEMPs) in central neurological disorders. *Clinical Neurophysiology.* 2016;127(1):40-49. doi:10.1016/j.clinph.2014.12.021
155. Rosengren SM, Colebatch JG, Straumann D, Weber KP. Why do oVEMPs become larger when you look up? Explaining the effect of gaze elevation on the ocular vestibular evoked myogenic potential. *Clinical Neurophysiology.* 2013;124(4):785-791. doi:10.1016/j.clinph.2012.10.012
156. Wirth MA, Valko Y, Rosengren SM, et al. Repetitive ocular vestibular evoked myogenic potential stimulation for the diagnosis of myasthenia gravis: Optimization of stimulation parameters. *Clinical Neurophysiology.* 2019;130(7):1125-1134. doi:10.1016/j.clinph.2019.03.033
157. Oh SJ. Distinguishing features of the repetitive nerve stimulation test between lambert-eaton myasthenic syndrome and myasthenia gravis, 50-year reappraisal. *J Clin Neuromuscul Dis.* 2017;19(2):66-75. doi:10.1097/CND.000000000000190
158. Reijntjes RH, Potters W V., Kerkhof FI, et al. Deriving reference values for nerve conduction studies from existing data using mixture model clustering. *Clinical Neurophysiology.* 2021;132(8):1820-1829. doi:10.1016/j.clinph.2021.04.013
159. de Meel RHP, Tannemaat MR, Verschuuren JJGM. Heterogeneity and shifts in distribution of muscle weakness in myasthenia gravis. *Neuromuscular Disorders.* 2019;29(9):664-670. doi:10.1016/j.nmd.2019.07.006

160. Fortin E, Cestari DM, Weinberg DH. Ocular myasthenia gravis: An update on diagnosis and treatment. *Curr Opin Ophthalmol*. 2018;29(6):477-484. doi:10.1097/ICU.0000000000000526
161. Grob D, Brunner N, Namba T, Pagala M. Lifetime course of myasthenia gravis. *Muscle Nerve*. 2008;37(2):141-149. doi:10.1002/mus.20950
162. Smith S V, Lee AG. Update on Ocular Myasthenia Gravis. *Neurol Clin*. 2017;35(1):115-123. doi:10.1016/j.ncl.2016.08.008
163. Claytor B, Li Y. Challenges in diagnosing coexisting ocular myasthenia gravis and thyroid eye disease. *Muscle Nerve*. 2021;63(5):631-639. doi:10.1002/mus.27118
164. De Meel RHP, Raadsheer WF, Van Zwet EW, Tannemaat MR, Verschuuren JJGM. Ocular Weakness in Myasthenia Gravis: Changes in Affected Muscles are a Distinct Clinical Feature. *J Neuromuscul Dis*. 2019;6(3):369-376. doi:10.3233/JND-190407
165. Walsh FB. Myasthenia Gravis and Its Ocular Signs: A Review. *Trans Am Ophthalmol Soc*. 1943;41:556.
166. Jellema HM, Braaksma-Besselink Y, Limpens J, Von Arx G, Wiersinga WM, Mourits MP. Proposal of success criteria for strabismus surgery in patients with Graves' orbitopathy based on a systematic literature review. *Acta Ophthalmol*. 2015;93(7):601-609. doi:10.1111/aos.12717
167. Dysli M, Fierz FC, Rappoport D, et al. Divergence bias in Hess compared to Harms screen strabismus testing. *Strabismus*. 2021;29(1):1-9. doi:10.1080/09273972.2020.1871382
168. Coll GE, Demer JL. The edrophonium-Hess screen test in the diagnosis of ocular myasthenia gravis. *Am J Ophthalmol*. 1992;114(4):489-493. doi:10.1016/S0002-9394(14)71863-X
169. Melcescu E, Horton WB, Kim D, et al. Graves Orbitopathy: Update on Diagnosis and Therapy. *South Med J*. 2014;107(1):34-43. doi:10.1097/SMJ.0000000000000038
170. McClelland C, Manousakis G, Lee MS. Progressive External Ophthalmoplegia. *Curr Neurol Neurosci Rep*. 2016;16(6):1-10. doi:10.1007/s11910-016-0652-7
171. Fischmann A. Oculopharyngeal muscular dystrophy. *Neuromuscular Imaging*. Published online January 1, 2013:305-311. doi:10.1007/978-1-4614-6552-2_24
172. Pineles SL, Rosenbaum AL, Demer JL. Changes in binocular alignment after surgery for concomitant and pattern intermittent exotropia. *Strabismus*. 2008;16(2):57-63. doi:10.1080/09273970802020292
173. Ben Simon GJ, Syed AM, Lee S, et al. Strabismus after Deep Lateral Wall Orbital Decompression in Thyroid-Related Orbitopathy Patients Using Automated Hess Screen. *Ophthalmology*. 2006;113(6):1050-1055. doi:10.1016/j.ophtha.2006.02.015
174. Flanders M. Restrictive strabismus: Diagnosis and management. *American Orthoptic Journal*. 2014;64(1):54-63. doi:10.3368/aoj.64.1.54
175. Aylward GW, McCarry B, Kousoulides L, Lee JP, Fells P. A scoring method for hess charts. *Eye (Basingstoke)*. 1992;6(6):659-661. doi:10.1038/eye.1992.141
176. Cogan DG, Yee RD, Gittinger J. Rapid eye movements in myasthenia gravis. I. Clinical observations. *Arch Ophthalmol*. 1976;94(7):1083-1085. doi:10.1001/ARCHOPHT.1976.03910040003001
177. Benatar M. A systematic review of diagnostic studies in myasthenia gravis. *Neuromuscular Disorders*. 2006;16(7):459-467. doi:10.1016/j.nmd.2006.05.006

178. O'hare M, Doughty C. Update on ocular myasthenia gravis. *Semin Neurol*. 2019;39(6):749-760. doi:10.1055/s-0039-1700527
179. de Meel RHP, Keene KR, Wirth MA, et al. Repetitive ocular vestibular evoked myogenic potentials in myasthenia gravis. *Neurology*. 2020;94(16):E1693-E1701. doi:10.1212/WNL.00000000000009306
180. Valko Y, Rosengren SM, Jung HH, Straumann D, Landau K, Weber KP. Ocular vestibular evoked myogenic potentials as a test for myasthenia gravis. *Neurology*. 2016;86(7):660-668. doi:10.1212/WNL.0000000000002383
181. Kocak GS, Tütüncü M, Adatepe NU, et al. A novel diagnostic method for myasthenia gravis. *Muscle Nerve*. 2021;64(3):328-335. doi:10.1002/mus.27353
182. Cleary M, Williams GJ, Metcalfe RA. The pattern of extra-ocular muscle involvement in ocular myasthenia. *Strabismus*. 2008;16(1):11-18. doi:10.1080/15569520701830992
183. Oosterhuis HJGH. The ocular signs and symptoms of myasthenia gravis. *Documenta Ophthalmologica*. 1982;52(2):363-378. doi:10.1007/BF01675867
184. Almog Y, Ben-David M, Nemet AY. Inferior oblique muscle paresis as a sign of myasthenia gravis. *Journal of Clinical Neuroscience*. 2016;25:50-53. doi:10.1016/j.jocn.2015.08.026
185. Akan O, Baysal-Kirac L. Ophthalmologic manifestations in myasthenia gravis: presentation and prognosis. *Acta Neurol Belg*. 2021;121(5):1131-1140. doi:10.1007/s13760-020-01556-3
186. McClelland C, Manousakis G, Lee MS. Progressive External Ophthalmoplegia. *Curr Neurol Neurosci Rep*. 2016;16(6). doi:10.1007/s11910-016-0652-7
187. Renard D, Ferraro A, Lorenzini MC, et al. Orthoptic and video-oculographic analyses in oculopharyngeal muscular dystrophy. *Muscle Nerve*. 2015;52(4):554-558. doi:10.1002/mus.24600
188. Shechtman D, Shallo-Hoffmann J, Rumsey J, Riordan-Eva P, Hardigan P. Maximum angle of ocular duction during visual fixation as a function of age. *Strabismus*. 2005;13(1):21-26. doi:10.1080/09273970590901810
189. Hering E. *The Theory of Binocular Vision*. New York: Plenum Press;; 1977.
190. Melson AT, McClelland CM, Lee MS. Ocular myasthenia gravis: Updates on an elusive target. *Curr Opin Neurol*. 2020;33(1):55-61. doi:10.1097/WCO.0000000000000775
191. Howard JF, Bril V, Vu T, et al. Safety, efficacy, and tolerability of efgartigimod in patients with generalised myasthenia gravis (ADAPT): a multicentre, randomised, placebo-controlled, phase 3 trial. *Lancet Neurol*. 2021;20(7):526-536. doi:10.1016/S1474-4422(21)00159-9
192. Europa TA, Nel M, Heckmann JM. A review of the histopathological findings in myasthenia gravis: Clues to the pathogenesis of treatment-resistance in extraocular muscles. *Neuromuscular Disorders*. 2019;29(5):381-387. doi:10.1016/j.nmd.2019.03.009
193. Fortin E, Cestari DM, Weinberg DH. Ocular myasthenia gravis: An update on diagnosis and treatment. *Curr Opin Ophthalmol*. 2018;29(6):477-484. doi:10.1097/ICU.0000000000000526
194. De Meel RHP, Raadsheer WF, Van Zwet EW, Tannemaat MR, Verschuuren JJGM. Ocular Weakness in Myasthenia Gravis: Changes in Affected Muscles are a Distinct Clinical Feature. *J Neuromuscul Dis*. 2019;6(3):369-376. doi:10.3233/JND-190407

195. Weber KP, Rappoport D, Dysli M, et al. Strabismus Measurements with Novel Video Goggles. *Ophthalmology*. 2017;124:1849-1856. doi:10.1016/j.ophtha.2017.06.020
196. Wary C, Azzabou N, Giraudeau C, et al. Quantitative NMRI and NMRS identify augmented disease progression after loss of ambulation in forearms of boys with Duchenne muscular dystrophy. *NMR Biomed*. 2015;28(9):1150-1162. doi:10.1002/nbm.3352
197. Willcocks RJ, Arpan IA, Forbes SC, et al. Longitudinal measurements of MRI-T2 in boys with Duchenne muscular dystrophy: Effects of age and disease progression. *Neuromuscular Disorders*. 2014;24(5):393-401. doi:10.1016/j.nmd.2013.12.012
198. Klupp E, Weidlich D, Schlaeger S, et al. B1-insensitive T2 mapping of healthy thigh muscles using a T2-prepared 3D TSE sequence. *PLoS One*. 2017;12(2):e0171337. doi:10.1371/journal.pone.0171337
199. Janiczek RL, Gambarota G, Sinclair CDJ, et al. Simultaneous T2 and lipid quantitation using IDEAL-CPMG. *Magn Reson Med*. 2011;66(5):1293-1302. doi:10.1002/mrm.22916
200. Forbes SC, Walter GA, Rooney WD, et al. Skeletal muscles of ambulant children with Duchenne muscular dystrophy: Validation of multicenter study of evaluation with MR imaging and MR spectroscopy. *Radiology*. 2013;269(1):198-207. doi:10.1148/radiol.13121948
201. Poon CS, Henkelman RM. Practical T2 quantitation for clinical applications. *Journal of Magnetic Resonance Imaging*. 1992;2(5):541-553. doi:10.1002/jmri.1880020512
202. Ben-Eliezer N, Sodickson DK, Block KT. Rapid and accurate T2 mapping from multi-spin-echo data using bloch-simulation-based reconstruction. *Magn Reson Med*. 2015;73(2):809-817. doi:10.1002/mrm.25156
203. Carlier PG. Global T2 versus water T2 in NMR imaging of fatty infiltrated muscles: Different methodology, different information and different implications. *Neuromuscular Disorders*. 2014;24(5):390-392. doi:10.1016/j.nmd.2014.02.009
204. Delfaut EM, Beltran J, Johnson G, Rousseau J, Marchandise X, Cotten A. Fat suppression in MR imaging: Techniques and pitfalls. *Radiographics*. 1999;19(2):373-382. doi:10.1148/radiographics.19.2.g99mr03373
205. Wokke BH, Van Den Bergen JC, Hooijmans MT, Verschuuren JJ, Niks EH, Kan HE. T2 relaxation times are increased in Skeletal muscle of DMD but not BMD patients. *Muscle Nerve*. 2016;53(1):38-43. doi:10.1002/mus.24679
206. Dula AN, Gochberg DF, Does MD. Optimal echo spacing for multi-echo imaging measurements of Bi-exponential T2 relaxation. *Journal of Magnetic Resonance*. 2009;196(2):149-156. doi:10.1016/j.jmr.2008.11.002
207. Azzabou N, De Sousa PL, Caldas E, Carlier PG. Validation of a generic approach to muscle water T2 determination at 3T in fat-infiltrated skeletal muscle. *Journal of Magnetic Resonance Imaging*. 2015;41(3):645-653. doi:10.1002/jmri.24613
208. Mankodi A, Azzabou N, Bulea T, et al. Skeletal muscle water T2 as a biomarker of disease status and exercise effects in patients with Duchenne muscular dystrophy. *Neuromuscular Disorders*. 2017;27(8):705-714. doi:10.1016/j.nmd.2017.04.008
209. McPhee KC, Wilman AH. Transverse relaxation and flip angle mapping: Evaluation of simultaneous and independent methods using multiple spin echoes. *Magn Reson Med*. 2017;77(5):2057-2065. doi:10.1002/mrm.26285

210. Lebel RM, Wilman AH. Transverse relaxometry with stimulated echo compensation. *Magn Reson Med*. 2010;64(4):1005-1014. doi:10.1002/mrm.22487
211. Weigel M. Extended phase graphs: Dephasing, RF pulses, and echoes - Pure and simple. *Journal of Magnetic Resonance Imaging*. 2015;41(2):266-295. doi:10.1002/jmri.24619
212. Kan HE, Scheenen TWJ, Wohlgemuth M, et al. Quantitative MR imaging of individual muscle involvement in facioscapulohumeral muscular dystrophy. *Neuromuscular Disorders*. 2009;19(5):357-362. doi:10.1016/j.nmd.2009.02.009
213. Yao L, Gai N. Fat-corrected T2 measurement as a marker of active muscle disease in inflammatory myopathy. *American Journal of Roentgenology*. 2012;198(5):W475-W481. doi:10.2214/AJR.11.7113
214. Stokes AM, Feng Y, Mitropoulos T, Warren WS. Enhanced refocusing of fat signals using optimized multipulse echo sequences. *Magn Reson Med*. 2013;69(4):1044-1055. doi:10.1002/mrm.24340
215. Froeling M, Hughes E, Schlawke L, Kan HE, Hollingsworth KG. The relation between fat calibration in multi-echo spin-echo water T2 mapping and STEAM fat T2 relaxation measurements. In: *Proceedings of the 27th Annual Meeting of ISMRM, Montréal, Canada*. ; 2019:1273.
216. Ren J, Dimitrov I, Sherry AD, Malloy CR. Composition of adipose tissue and marrow fat in humans by 1H NMR at 7 Tesla. *J Lipid Res*. 2008;49(9):2055-2062. doi:10.1194/jlr.D800010-JLR200
217. Hooijmans MT, Baligand C, Froeling M, et al. Multi-parametric {MR} shows increased T2 heterogeneity in fat infiltrated muscles in Becker Muscular Dystrophy. In: *Proc. Joint Annual Meeting ISMRM-ESMRMB, Paris, France*. ; 2018:816.
218. Otto LAM, Froeling M, Van Den Berg LH, Hendrikse J, van der Pol WL. Muscle MRI in a cross-sectional cohort of patients with Spinal Muscular Atrophy types 2-3. In: *Journal of Neuromuscular Diseases*. Vol 6. ; 2019:S54-S55.
219. Hooijmans MT, Niks EH, Burakiewicz J, et al. Non-uniform muscle fat replacement along the proximodistal axis in Duchenne muscular dystrophy. *Neuromuscular Disorders*. 2017;27(5):458-464. doi:10.1016/j.nmd.2017.02.009
220. Naarding K, Veeger T, Sardjoe Mishre A, et al. P.305MRI brachialis contractile cross-sectional area is correlated strongest to elbow flexion in non-ambulant Duchenne muscular dystrophy patients. In: *Neuromuscular Disorders*. Vol 29. ; 2019:S156. doi:10.1016/j.nmd.2019.06.419
221. Thomas LW. the Chemical Composition of Adipose Tissue of Man and Mice. *Q J Exp Physiol Cogn Med Sci*. 1962;47(2):179-188. doi:10.1113/expphysiol.1962.sp001589
222. Schlaeger S, Weidlich D, Klupp E, et al. Decreased water T2 in fatty infiltrated skeletal muscles of patients with neuromuscular diseases. *NMR Biomed*. 2019;32(8):e4111. doi:10.1002/nbm.4111
223. Arnold WD, Kassam D, Kissel JT. Spinal muscular atrophy: Diagnosis and management in a new therapeutic era. *Muscle Nerve*. 2015;51(2):157-167. doi:10.1002/mus.24497
224. Desguerre I, Mayer M, Leturcq F, Barbet JP, Gherardi RK, Christov C. Endomysial fibrosis in duchenne muscular dystrophy: A marker of poor outcome associated with macrophage alternative activation. *J Neuropathol Exp Neurol*. 2009;68(7):762-773. doi:10.1097/NEN.0b013e3181aa31c2

225. Saab G, Thompson RT, Marsh GD. Multicomponent T2 relaxation of in vivo skeletal muscle. *Magn Reson Med*. 1999;42(1):150-157. doi:10.1002/(SICI)1522-2594(199907)42:1<150::AID-MRM20>3.0.CO;2-5
226. Querleux B, Cornillon C, Jolivet O, Bittoun J. Anatomy and physiology of subcutaneous adipose tissue by in vivo magnetic resonance imaging and spectroscopy: Relationships with sex and presence of cellulite. *Skin Research and Technology*. 2002;8(2):118-124. doi:10.1034/j.1600-0846.2002.00331.x
227. Beekman R, Kuks JBM, Oosterhuis HJGH. Myasthenia gravis: Diagnosis and follow-up of 100 consecutive patients. *J Neurol*. 1997;244(2):112-118. doi:10.1007/s004150050059
228. Mazzoli M, Ariatti A, Valzania F, et al. Factors affecting outcome in ocular myasthenia gravis. *International Journal of Neuroscience*. 2018;128(1):15-24. doi:10.1080/00207454.2017.1344237
229. J. Smith T. TSHR as a therapeutic target in Graves' disease. *Expert Opin Ther Targets*. 2017;21(4):427-432. doi:10.1080/14728222.2017.1288215
230. Kung FP, Hung HC, Ou HY, Wu TJ. Graves' ophthalmopathy. *Journal of Internal Medicine of Taiwan*. 2012;23(1):9-20. doi:10.5005/jp/books/10258_7
231. De Abreu MR, Chung CB, Biswal S, Haghighi P, Hesselink J, Resnick D. Erdheim-Chester Disease: MR Imaging, Anatomic, and Histopathologic Correlation of Orbital Involvement. *American Journal of Neuroradiology*. 2004;25(4):627-630.
232. Das T, Roos JCP, Patterson AJ, Graves MJ, Murthy R. T2-relaxation mapping and fat fraction assessment to objectively quantify clinical activity in thyroid eye disease: an initial feasibility study. *Eye (Basingstoke)*. 2019;33(2):235-243. doi:10.1038/s41433-018-0304-z
233. Han E, Gold G, Stainsby J, Beaulieu J, Brittain J. *In-Vivo T1 and T2 Measurements of Muskuloskeletal Tussie at 3T and 1.5T*. Vol 166.; 2003.
234. Yushkevich PA, Piven J, Hazlett HC, et al. User-guided 3D active contour segmentation of anatomical structures: Significantly improved efficiency and reliability. *Neuroimage*. 2006;31(3):1116-1128. doi:10.1016/j.neuroimage.2006.01.015
235. Keene KR, Beenakker JWM, Hooijmans MT, et al. T2 relaxation-time mapping in healthy and diseased skeletal muscle using extended phase graph algorithms. *Magn Reson Med*. 2020;84(5):2656-2670. doi:10.1002/mrm.28290
236. Mourits MP, Prummel MF, Wiersinga WM, Koornneef L. Clinical activity score as a guide in the management of patients with Graves' ophthalmopathy. *Clin Endocrinol (Oxf)*. 1997;47(1):9-14. doi:10.1046/j.1365-2265.1997.2331047.x
237. Kiefer LS, Fabian J, Lorbeer R, et al. Inter- and intra-observer variability of an anatomical landmark-based, manual segmentation method by MRI for the assessment of skeletal muscle fat content and area in subjects from the general population. *British Journal of Radiology*. 2018;91(1089). doi:10.1259/bjr.20180019
238. Hiba B, Richard N, Hébert LJ, et al. Quantitative assessment of skeletal muscle degeneration in patients with myotonic dystrophy type 1 using MRI. *Journal of Magnetic Resonance Imaging*. 2012;35(3):678-685. doi:10.1002/jmri.22849
239. Hernando D, Sharma SD, Aliyari Ghasabeh M, et al. Multisite, multivendor validation of the accuracy and reproducibility of proton-density fat-fraction quantification at 1.5T and 3T using a fat-water phantom. *Magn Reson Med*. 2017;77(4):1516-1524. doi:10.1002/mrm.26228

240. Morrow JM, Sinclair CDJ, Fischmann A, et al. Reproducibility, and age, body-weight and gender dependency of candidate skeletal muscle MRI outcome measures in healthy volunteers. *Eur Radiol.* 2014;24(7):1610-1620. doi:10.1007/s00330-014-3145-6
241. Schlaffke L, Rehmann R, Rohm M, et al. Multi-center evaluation of stability and reproducibility of quantitative MRI measures in healthy calf muscles. *NMR Biomed.* 2019;32(9):e4119. doi:10.1002/nbm.4119
242. Hu HH, Kan HE. Quantitative proton MR techniques for measuring fat. *NMR Biomed.* 2013;26(12):1609-1629. doi:10.1002/nbm.3025
243. Liu CY, McKenzie CA, Yu H, Brittain JH, Reeder SB. Fat quantification with IDEAL gradient echo imaging: Correction of bias from T1 and noise. *Magn Reson Med.* 2007;58(2):354-364. doi:10.1002/mrm.21301
244. Forbes SC, Arora H, Willcocks RJ, et al. Upper and lower extremities in Duchenne muscular dystrophy evaluated with quantitative MRI and proton MR spectroscopy in a multicenter cohort. *Radiology.* 2020;295(3):616-625. doi:10.1148/radiol.2020192210
245. Higashiyama T, Iwasa M, Ohji M. Quantitative Analysis of Inflammation in Orbital Fat of Thyroid-associated Ophthalmopathy Using MRI Signal Intensity. *Sci Rep.* 2017;7(1). doi:10.1038/s41598-017-17257-6
246. Cros D, Harnden P, Pellissier JF, Serratrice G. Muscle hypertrophy in Duchenne muscular dystrophy - A pathological and morphometric study. *J Neurol.* 1989;236(1):43-47. doi:10.1007/BF00314217
247. Rautenbach RM, Pillay K, Murray ADN, Heckmann JM. Extraocular muscle findings in myasthenia gravis associated treatment-resistant ophthalmoplegia. *Journal of Neuro-Ophthalmology.* 2017;37(4):414-417. doi:10.1097/WNO.0000000000000534
248. Moon SY, Lee SS, Hong YH. Muscle atrophy in muscle-specific tyrosine kinase (MuSK)-related myasthenia gravis. *Journal of Clinical Neuroscience.* 2011;18(9):1274-1275. doi:10.1016/j.jocn.2011.01.010
249. Paz ML, Barrantes FJ. Autoimmune Attack of the Neuromuscular Junction in Myasthenia Gravis: Nicotinic Acetylcholine Receptors and Other Targets. *ACS Chem Neurosci.* 2019;10(5):2186-2194. doi:10.1021/acscchemneuro.9b00041
250. Walters J. Muscle hypertrophy and pseudohypertrophy. *Pract Neurol.* 2017;17(5):369-379. doi:10.1136/practneurol-2017-001695
251. Tian S, Nishida Y, Isberg B, Lennerstrand G. MRI measurements of normal extraocular muscles and other orbital structures. *Graefe's Archive for Clinical and Experimental Ophthalmology.* 2000;238(5):393-404. doi:10.1007/s004170050370
252. Wong SH. Advocating Patient-Centred Research in Ocular Myasthenia Gravis (OMG): A Call for an OMG Research Consortium. *Frontiers in Ophthalmology.* 2022;0:35. doi:10.3389/FOPHT.2022.912805
253. Keene KR, de Nie JM, Brink MJ, et al. Diagnosing myasthenia gravis using orthoptic measurements: assessing extraocular muscle fatiguability. *J Neurol Neurosurg Psychiatry.* 2022;0:jnnp-2022-329859. doi:10.1136/JNNP-2022-329859
254. Nehrke K, Versluis MJ, Webb A, Börner P. Volumetric B1 (+) mapping of the brain at 7T using DREAM. *Magn Reson Med.* 2014;71(1):246-256. doi:10.1002/MRM.24667
255. Bruenech JR, Kjellevoid Haugen IB. How does the structure of extraocular muscles and their nerves affect their function? In: *Eye (Basingstoke).* Vol 29. Nature Publishing Group; 2015:177-183. doi:10.1038/eye.2014.269

256. Spencer RF, Porter JD. Structural organization of the extraocular muscles. *Rev Oculomot Res.* 1988;2:33-79. Accessed February 15, 2021. <https://pubmed.ncbi.nlm.nih.gov/3153651/>
257. Haładaj R. Comparison of lateral and medial rectus muscle in human: an anatomical study with particular emphasis on morphology, intramuscular innervation pattern variations and discussion on clinical significance. *Surgical and Radiologic Anatomy.* 2020;42(5):607-616. doi:10.1007/s00276-019-02400-x
258. Ironside R. Progressive Exophthalmoplegia with Muscular Atrophy, Myasthenia, and Thyrotoxicosis. *J R Soc Med.* 1951;44(8):690-691. doi:10.1177/003591575104400812
259. Kornegay JN, Childers MK, Bogan DJ, et al. The Paradox of Muscle Hypertrophy in Muscular Dystrophy. *Phys Med Rehabil Clin N Am.* 2012;23(1):149-172. doi:10.1016/j.pmr.2011.11.014
260. Krampitz DE, Wolfe GI, Fleckenstein JL, Barohn RJ. Charcot-Marie-Tooth disease type 1A presenting as calf hypertrophy and muscle cramps. *Neurology.* 1998;51(5):1508-1509. doi:10.1212/WNL.51.5.1508
261. Jaarsma-Coes MG, Ferreira TA, van Houdt PJ, van der Heide UA, Luyten GPM, Beenakker JWM. Eye-specific quantitative dynamic contrast-enhanced MRI analysis for patients with intraocular masses. *MAGMA.* 2022;35(2):311-323. doi:10.1007/S10334-021-00961-W
262. Finlayson S, Morrow JM, Rodriguez Cruz PM, et al. Muscle magnetic resonance imaging in congenital myasthenic syndromes. *Muscle Nerve.* 2016;54(2):211-219. doi:10.1002/mus.25035
263. Demer JL. The orbital pulley system: A revolution in concepts of orbital anatomy. *Ann N Y Acad Sci.* 2002;956(1):17-32. doi:10.1111/j.1749-6632.2002.tb02805.x
264. Spencer RF, Porter JD. Biological organization of the extraocular muscles. *Prog Brain Res.* 2006;151:43-80. doi:10.1016/S0079-6123(05)51002-1
265. Kirchner J, Watson T, Lappe M. Real-Time MRI Reveals Unique Insight into the Full Kinematics of Eye Movements. *eNeuro.* 2022;9(1). doi:10.1523/ENEURO.0357-21.2021
266. Heskamp L, Miller AR, Birkbeck MG, et al. In vivo 3D imaging of human motor units in upper and lower limb muscles. *Clinical Neurophysiology.* Published online June 20, 2022. doi:10.1016/j.clinph.2022.05.018
267. Birkbeck MG, Heskamp L, Schofield IS, Blamire AM, Whittaker RG. Non-invasive imaging of single human motor units. *Clinical Neurophysiology.* 2020;131(6):1399-1406. doi:10.1016/j.clinph.2020.02.004
268. Heskamp L, Birkbeck MG, Whittaker RG, Schofield IS, Blamire AM. The muscle twitch profile assessed with motor unit magnetic resonance imaging. *NMR Biomed.* 2021;34(3). doi:10.1002/nbm.4466

