Medscape Education

Digital Medical Simulation Platform Improves Neurologists' Ability to Assess for and Diagnose Muscular Dystrophies

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BACKGROUND

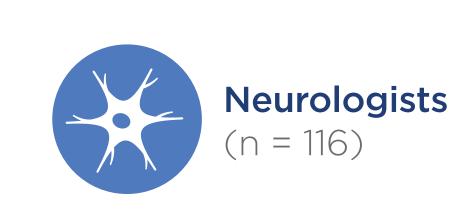
Duchenne muscular dystrophy (DMD) has a prevalence of 4.8/100,000 people and the prevalence of limb-girdle muscular dystrophy (LGMD) is estimated to be between 1/14,500 and 1/123,000 people.^{1,2} Symptom burden is substantial for both conditions for which timely access to therapeutic modalities is recommended. Access to

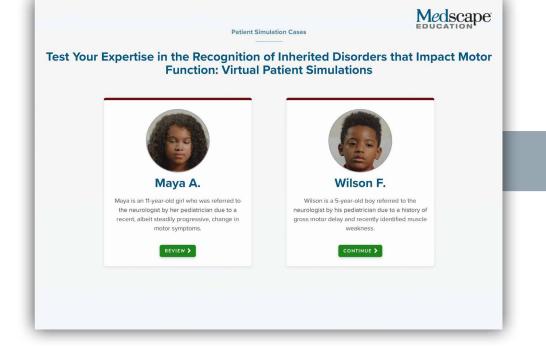


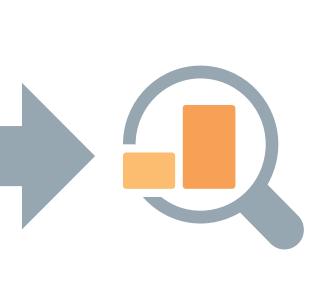
treatment may be hindered by data showing that both forms of muscular dystrophy are associated with a delay in diagnosis.^{3,4} To help address the delay in diagnosis, this study utilized an online medical simulation platform to improve the ability of neurologists' to assess, differentiate between, and diagnose either DMD or LGMD.

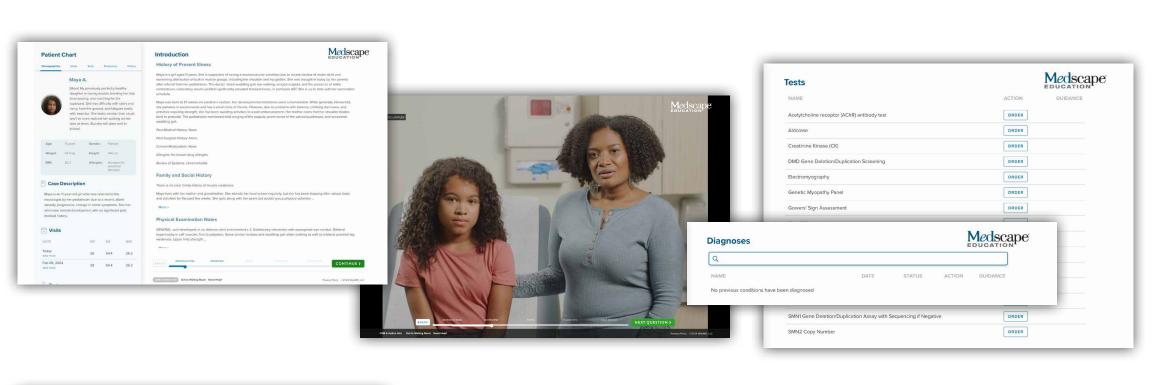
METHODS

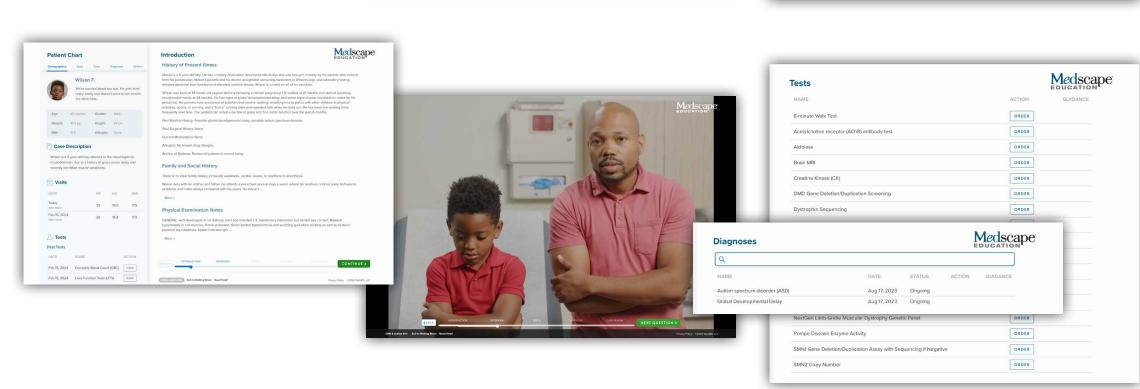
The intervention comprised 2 patients presenting in a virtual patient simulation (VPS) platform that allows learners to order clinical tests and make a diagnosis in a manner matching the scope and depth of actual practice. Tailored clinical guidance (CG), based on current evidence and expert recommendation, was provided following each decision, followed by the opportunity for the learner to modify their decisions. Decisions were collected post-CG and compared with each user's baseline (pre-CG) decisions using a McNemar's test to determine P values. Data were collected from September 2022 through Decmber 2022.5











RESULTS

PATIENT CASE 1: ASSESSMENT AND DIAGNOSIS OF LGMD



Neurologists improved their ability to select appropriate tests for a patient who presents with MAYA recent progressive decline in motor function

Order: Gowers' Sign

Order: Genetic Myopathy Panel

Order: Creatinine Kinase (CK)

diagnosing LGMD

CASE SUMMARY

Maya is a girl aged 11 years. She is suspected of having a neuromuscular condition due to recent decline of motor skills and worsening distribution of bulk in muscle groups, including the shoulder and hip girdles. She was brought in today by her parents after referral from her pediatrician. The doctor noted waddling gait, toe-walking, winged scapula, and the presence of ankle contractures. Laboratory results yielded significantly elevated transaminases, in particular AST. She is up to date with her vaccination schedule.

Maya was born at 37 weeks via elective c-section. Her developmental milestones were unremarkable. While generally introverted, she partakes in social events and has a small circle of friends. However, due to problems with balance, climbing staircases, and activities requiring strength, she has been avoiding activities to avoid embarrassment. Her mother notes that her shoulder blades tend to protrude. The pediatrician mentioned mild winging of the scapula, prominence of the calves/quadriceps, and occasional waddling gait.

Allergies: Nonspecific seasonal

Muscular Dystrophy Type 2E (LGMD2E Muscular Dystrophy

38% **43**% **15**%

Selecting appropriate assessment tools

for pediatric patients who present with

■ PRE GUIDANCE
■ POST GUIDANCE
■ % POINT CHANGE

Making an accurate diagnosis in a

timely manner that explains the initial

symptoms of weakness and impaired

Neurologists demonstrate improvement in

54% 76% †22%

89% 97<mark>% 18% NS</mark>

76% 89% ↑13% *P* <.05

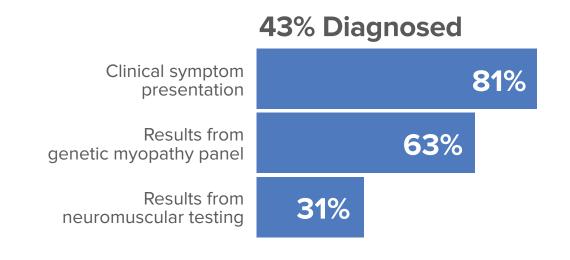
P <.01

symptoms of weakness and impaired

P < .05

Clinicians identify what influenced their diagnostic decision

Why Did Neurologists Diagnose Limb Girdle Muscular Dystrophy (LGMD)?



57% Did Not Diagnose The symptom presentation formed the basis of my diagnosis The results of neuromuscular testing formed the basis of my diagnosis I'm not sure how to assess a patient with these symptoms

Why Did Neurologists Diagnose Limb Girdle Muscular Dystrophy Type 2E (LGMD2E)?

	54% Diagnosed	
Results from genetic myopathy panel		75 %
Clinical symptom presentation		70%
Results from neuromuscular testing	30%	

46% Did Not Diagnose The symptom presentation formed the basis of my diagnosis I'm not sure how to assess a patient with these symptoms The results of neuromuscular testing formed the basis of my diagnosis The results of genetic testing formed the basis of my diagnosis

PATIENT CASE 2: ASSESSMENT AND DIAGNOSIS OF DMD



WILSON CASE SUMMARY

Wilson is a 5-year-old boy. He has a history of possible developmental delays and was brought in today by his parents after referral from his pediatrician. Wilson's parents and his doctor recognized worsening weakness in Wilson's legs, and laboratory testing showed abnormal liver function and elevated creatine kinase. Wilson is

Wilson was born at 38 weeks via vaginal delivery following a normal pregnancy. He walked at 21 months and started speaking recognizable words at 28 months. He has signs of global developmental delay, and some signs of poor socialization noted by his preschool. His parents note avoidance of activities that involve walking; unwillingness to join in with other children in physical activities, sports, or running; and a "funny" running style and repeated falls when he does run. He has been toe walking more frequently over time. The pediatrician noted a decline in gross and fine motor function over the past 6 months.

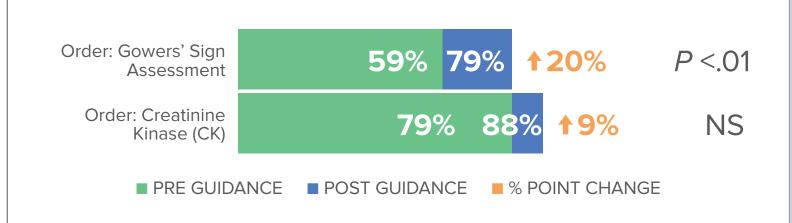
Gender: Male

current on all of his vaccines.

Weight: 16.6 kg

Assessment of patients who may be experiencing seizures

Neurologists improved their ability to select appropriate tests for a patient with gross motor delay and muscle weakness



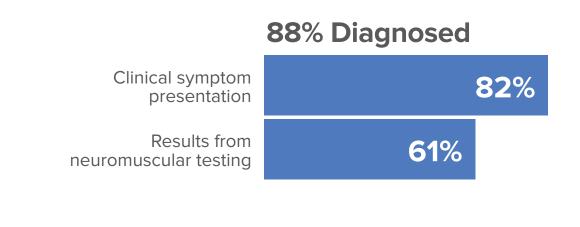
Making an accurate diagnosis in a timely manner that explains the initial symptoms of weakness and impaired

Neurologists demonstrate improvement in diagnosing DMD



Clinicians identify what influenced their diagnostic decision

Why Did Neurologists Diagnose Duchenne Muscular Dystrophy?



12% Did Not Diagnose I'm not sure how to assess a patient with these symptoms The symptom presentation formed the basis of my The results of neuromuscular testing formed the basis of my diagnosis







CONCLUSIONS

This online CME-certified medical simulation program was effective in improving the assessment and diagnosis of patients with muscular dystrophies. In the first case of an 11-year-old patient, almost every learner chose to perform genetic testing specific to myopathies. The education was most successful in improving the use of Gower's sign assessment, which is a characteristic neurologic assessment specific to DMD. Armed with test results, nearly 80% of neurologists correctly diagnosed the patient with LGMD or a more specific diagnosis of LGMD type 2. Despite genetic testing providing definitive cause of the symptoms, most people stated that the patient's clinical symptoms contributed the most to making the diagnosis. Neuromuscular testing was noted to be the least helpful test modality for this patient. Many of those who did not make a correct diagnosis relied on results from neuromuscular testing.

In the second case, which was of a 5-year-old patient presenting with gross motor delay and muscle weakness, most neurologists correctly chose to measure creatinine kinase levels and assess Gowers' sign. Unlike the LGMD case, less than two-third of neurologists correctly diagnosed the patient with DMD. CG resulted in a 26% relative improvement in neurologists making the correct diagnosis. Clinical symptom presentation was a key diagnostic tool for almost all neurologists who made a correct diagnosis. Among those who did not correctly diagnose the patient, the nearly 60% reported that they were unsure of how to assess the patient.

The results of this online medical simulation support the need for additional education for diagnosing muscular dystrophies. Neurologists would benefit from additional education on the signs and symptoms of specific muscular dystrophies, key neurologic assessments and genetic testing to narrow down the diagnosis, and a review of the diagnostic criteria for specific muscular dystrophies.

ACKNOWLEDGEMENTS

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