

MAG-Associated Demyelinating Polyneuropathy: A Case Study and Review



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Introduction

- A 40 year old woman presented with atrophy and loss of function in her hands. In the past, she had seen primary care providers who thought her symptoms were caused by compression and possible carpal tunnel syndrome. In 2021, the patient underwent carpal tunnel surgery for treatment of the left wrist. However, following the procedure not only did her symptoms not resolve, but they progressively started getting worse. She was then referred to neurology for further workup. The neurologist conducted a thorough physical exam as well as ordered an US, EMG, blood work, lumbar puncture, and an autoimmune panel. When combining her physical exam, labs, and imaging findings, the neurologist was able to conclude that the patient likely had MAG associated polyneuropathy.
- MAG Associated Demyelinating Polyneuropathy is a rare autoimmune condition where IgM in the host mistakenly attacks its own myelin associated glycoprotein (MAG), resulting in the loss of function in peripheral nerves. The condition is very rare, with only 0.8 to 8.9 new cases found per 100,000 per year.¹

References

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2. Rosario, N. B., & De Jesus, O. (2023). *Electrodiagnostic evaluation of carpal tunnel syndrome*. In StatPearls. StatPearls Publishing. <https://www.ncbi.nlm.nih.gov/books/NBK562235/>
3. Steck, A. J., et al. (2021). Anti-MAG neuropathy: From biology to clinical management. *Journal of Neuroimmunology*, 361, 577726. <https://doi.org/10.1016/j.jneuroim.2021.577726>
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Methodology

The patient underwent electrodiagnostic testing to look for any abnormalities in motor, sensory, and F-wave values.

Motor NCS

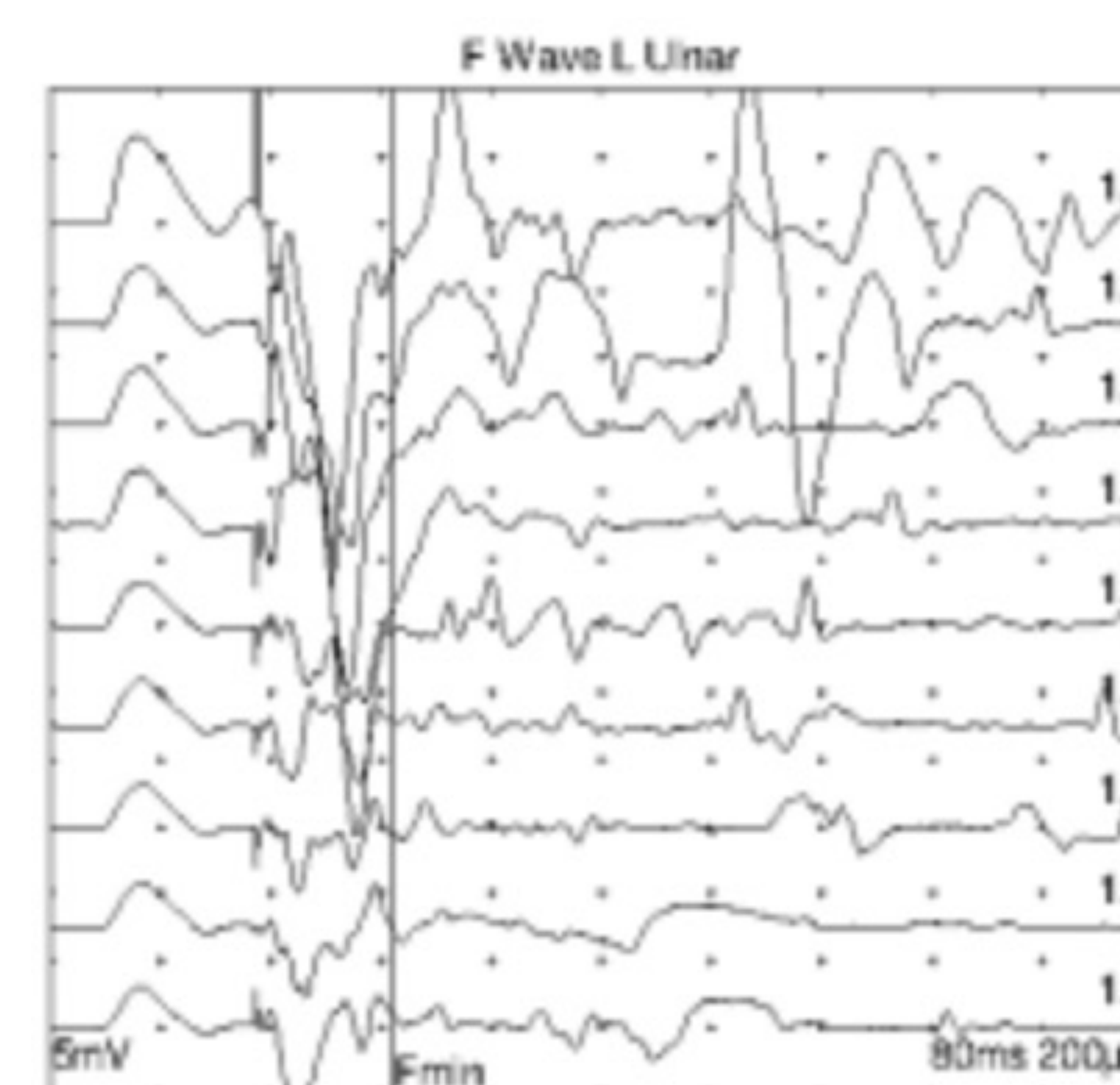
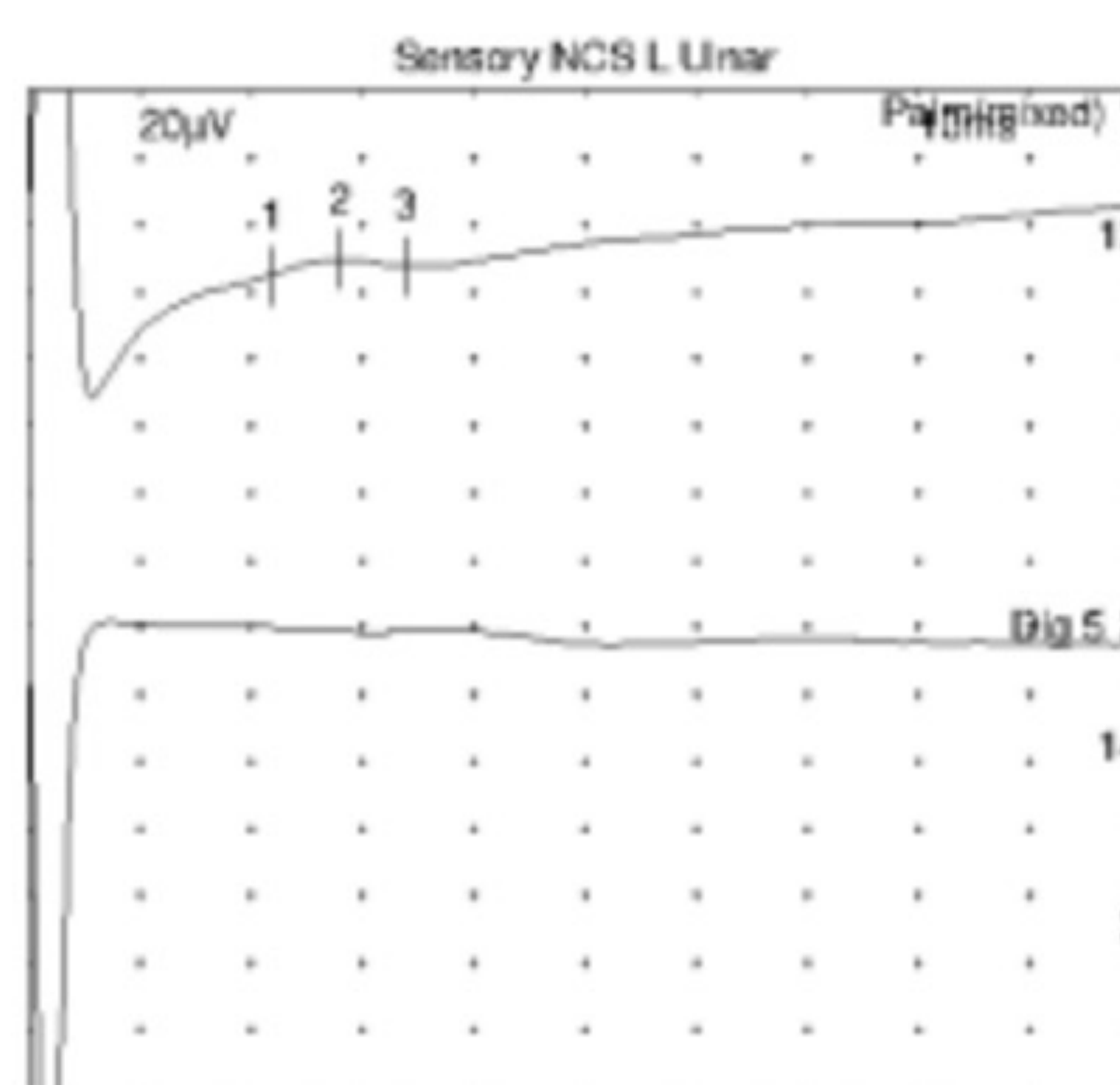
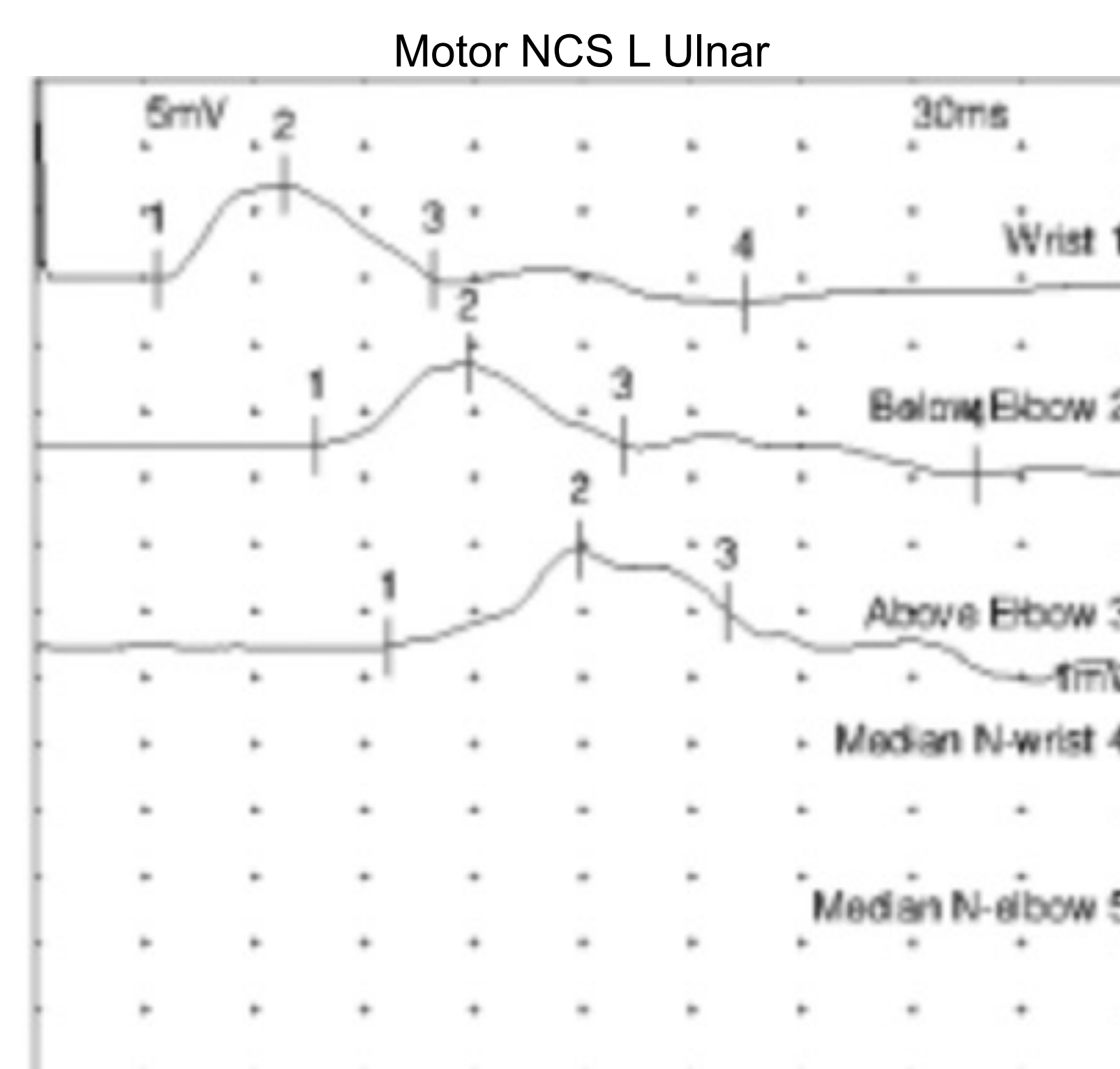
Nerve / Sites	Lat. ms	Amp. mV	Dist. cm	Vel. m/s	Temp. °C
R Median - APB					
Wrist	3.91	9.6	6.5		36.3
Ref.	4.40	4.4			
Elbow	10.83	1.4	24.5	35.4	36.3
Ref.		4.4		49.0	
L Median - APB					
Wrist	2.86	11.4	6.5		36.5
Ref.	4.40	4.4			
Elbow	7.29	8.9	23	52.0	36.5
Ref.		4.4		49.0	
R Ulnar - ADM					
Wrist	2.45	11.4	6.5		36.3
Ref.	3.50	5.6			
Below Elbow	9.58	0.3	22	30.8	36.3
Ref.		5.6		49.0	
Above Elbow	15.05	1.5	7	12.8	36.3
Ref.		5.6		49.0	
Median N-wrist	NR	NR			
Median N-elbow	NR	NR			
L Ulnar - ADM					
Wrist	3.33	7.0	6.5		36.5
Ref.	3.50	5.6			
Below Elbow	7.71	6.0	22	50.3	36.5
Ref.		5.6		49.0	
Above Elbow	9.69	1.5	8	40.4	
Ref.		5.6		49.0	
R Peroneal - EDB					
Ankle	4.27	3.1	7.5		34.7
Ref.	5.70	2.2			
Fib Head	11.15	2.8	32	46.5	34.7
Ref.		2.2		41.0	
Pop Fossa	12.66	2.8	8	53.0	34.7
Ref.		2.2		41.0	
R Tibial - AH					
Ankle	4.01	21.0	7.5		34.7
Ref.	5.70	2.5			
Pop Fossa	11.82	11.1	38	48.6	34.7
Ref.		2.5		39.0	

Sensory NCS

Nerve / Sites	Rec. Site	Peak Lat ms	Ampl. µV	Dist. cm	Vel. m/s	Temp. °C
R Median						
Palm(mixed)	Wrist	1.88	76.8	8	56.9	36.3
Ref.		2.20	40.0			
Dig 2	Wrist	2.76	25.9	13	60.9	36.3
Ref.		3.50	10.0			
L Median						
Palm(mixed)	Wrist	1.77	25.9	8	61.4	36.5
Ref.		2.20	40.0			
Dig 2	Wrist	2.34	9.6	13	71.3	36.5
Ref.		3.50	10.0			
R Ulnar						
Palm(mixed)	Wrist	1.82	9.1	8	59.1	36.3
Ref.		2.20	20.0			
Dig 5	Wrist	2.34	3.1	11	58.7	36.3
Ref.		2.90	5.0			
L Ulnar						
Palm(mixed)	Wrist	2.81	4.7	8	36.6	36.5
Ref.		2.20	20.0			
Dig 5	Wrist	NR	NR	11	NR	
Ref.		2.90	5.0			
L Radial						
Forearm	Snuffbox	1.93	58.3	10	68.6	36.5
Ref.		2.60	25.0			

F Wave

Nerve	F min ms
R Median	36.88
R Ulnar	30.73
R Peroneal	65.57
R Tibial	51.46
L Median	26.41
L Ulnar	24.79



Results

- Currently, the gold standard for diagnosis of symptomatic carpal tunnel syndrome is electrodiagnostic studies.² However, her EMG and US showed she had multiple sites of conduction blockage at noncompressible sites, supporting the conclusion that her symptoms were not due to a compressive etiology such as carpal tunnel syndrome.
- The patient's MAG IGM autoantibody panel came back positive with a high value of 5725 btu.
- More specifically, the patient displayed conduction blocks at these specific sites: proximal right median and right ulnar nerves in the area between the elbow and wrist, another in the right ulnar nerve near the elbow, and one in the left ulnar nerve near the elbow
- As seen in the figures, nerve blockage and temporal dispersion are seen in the L ulnar nerve indicating possible demyelination and CIDP
- The F-Wave of the left ulnar nerve has a lower than normal value of 24.79 (Normal: 25-32 ms in UE).⁴

Conclusion

- In this case, the patient underwent carpal tunnel surgery of the left wrist due to a misdiagnosis. Only through EMG and US was the diagnosis able to be found.
- Once diagnosed with MAG associated polyneuropathy, the patient was able to be started on rituximab and has since seen improvement.
- With severe symptoms such as weakness or sensory deficit, these patients should undergo confirmatory nerve conduction studies before any invasive procedures are done.
- Current recommended treatment for MAG polyneuropathy is rituximab but development of improved treatments are underway.³
- Further study on the condition is necessary to improve screening algorithms and to improve the care of affected patients in the future.