Database Evaluation for Muscle and Nerve Diseases – DEMAND: An academic neuromuscular coding system

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ABSTRACT

Background: A database which documents the diagnosis of neuromuscular patients is useful for determining the types of patients referred to academic centers and for identifying participants for clinical trials and other studies. The ICD-9 or ICD-10 numeric systems are insufficiently detailed for this purpose.

Objective: To develop a database for neuromuscular diagnoses

Methods: We developed a detailed diagnostic coding system for neuromuscular diseases called DEMAND: Database Evaluation for Muscle and Nerve Diseases that has been adopted by neuromuscular clinics at University of Texas Health Science Center San Antonio (UTHSCSA), Ohio State University (OSU), University of Kansas Medical Center (KUMC), and University of Texas Southwestern (UTSW). At the initial visit, patients are assigned a diagnostic code which can be revised later if appropriate. Fields include patient's name, date of birth, and diagnostic code. The neuromuscular database consisted of 457 codes. Each code has a prefix (MUS or PNS) followed by a three-digit number. Depending on whether muscle or nerve is primarily involved, there are eight broad groups: motor neuron disease (MUS codes 100-139); neuromuscular junction disorders (MUS 200-217); acquired and hereditary myopathies (MUS 300-600s); acquired and hereditary polyneuropathies (PNS 100-400); mononeuropathies (PNS 500s); plexopathies (PNS 600s); radiculopathies (PNS 700s); and mononeuritis multiplex (PNS 800s).

Results: During a period of 10 years, 17,163 of patients were entered (1,752 at UTHSCSA, 1,840 at OSU, 3,699 at KUMC, 9,872 at UTSW). The number of patients in several broad categories are: 3,080 motor neuron disease; 1,575 neuromuscular junction disease; 1,851 muscular dystrophies; 633 inflammatory myopathies; 1,090 hereditary neuropathies; 1,001 immune-mediated polyneuropathies; 620 metabolic/toxic polyneuropathies; 535 mononeuropathies; 296 plexopathies; and 769 radiculopathies.

Conclusion: A detailed diagnostic neuromuscular database can be utilized at multiple academic centers. The database should be simple without too many fields to complete, to ensure compliance during busy clinic operations. This database has been very useful in identifying groups of patients for retrospective, observational studies and for prospective treatment studies including trials for Amyotrophic Lateral Sclerosis (ALS), Muscular Dystrophies (MD), Myasthenia Gravis (MG), and retrospective studies of Primary Lateral Sclerosis (PLS), chronic inflammatory demyelinating neuropathy (CIDP), etc.

Introduction

Even when the ICD-9 transitioned to ICD-10, there was still not enough precision for academic purposes. The ICD system is primarily a billing system and not an academic classification system. A database which documents the diagnosis of neuromuscular patients is valuable for determining the types of patients referred to academic centers and for identifying participants for future studies and clinical trials. The ICD-9 or ICD-10 numeric system is poorly suited for this purpose. For instance, all forms of muscular dystrophy and hereditary neuropathy are assigned one code in the ICD-9 or ICD-10 system (Myotonic dystrophy type 1 and Myotonic dystrophy type 2 are assigned the same codes in ICD-9 or ICD-10 system). Due to the wide variety of different muscle disorders and the myriad etiologies for polyneuropathies, the neuromuscular database is poorly served by the ICD-9 or ICD-10. In addition, the ICD-9 and ICD-10 system may encounter difficulty incorporating new genetic and other diagnoses as they are recognized over time. We developed a database for neuromuscular diagnoses. Our goal was to develop a comprehensive uniform database to diagnose neuromuscular patients for all study designs which can also be adapted to allow mapping to new nomenclature and reclassification. We have called this coding system DEMAND: Database Evaluation for Muscle and Nerve Disease.

Methods

We developed a comprehensive diagnostic coding system for neuromuscular diseases. The system was first developed at University of Texas Southwestern (UTSW) in 1993. It was later adopted at University of Texas Health Science Center San Antonio (UTHSCSA), Ohio State University (OSU), and University of Kansas Medical Center (KUMC). The neuromuscular database initially consisted of fewer codes but over time has expanded to 457 codes. Each code has a prefix (MUS or PNS) followed by a three-digit number. There are eight broad groups:

Motor Neuron Disease	MUS 100-139
Neuromuscular Junction	MUS 200-217
Acquired and Hereditary Myopathies	MUS 300-600
Acquired and Hereditary Polyneuropathies	PNS 100-400
Mononeuropathies	PNS 500
Plexopathies	PNS 600
Radiculopathies	PNS 700
Mononeuritis Multiplex	PNS 800

For reference purposes, the corresponding ICD-10 and ICD-9 numbers for each MUS and PNS code are listed. For details regarding all the individual codes in each broad group along with the corresponding ICD-10 and ICD-9 numbers, see Appendix 1 for each MUS code and Appendix 2 for each PNS code. At the time of collection of this data, only the ICD-9 codes were available. However, since then the ICD-10 codes have also been released. If a disease diagnosis is not apparent, the database allows the option by symptoms or lab findings. Examples of symptoms are Fatigue – MUS 811, Numbness – PNS 835, Myalgias – MUS 301, and Pain – PNS 820.

Neuromuscular Clinical Database Collection form (Appendix 3)

The neuromuscular clinical database contains patient's names, ID, date of birth, social security number, neuromuscular code (MUS or PNS), ICD code, date of visit, and new or follow-up visit. We ask physicians to complete these after they see the patient. The information is then transferred to a computerized spread sheet at each site. With the introduction of the electronic health record some sites determined ways to enter the PNS/MUS code directly in the electronic medical record. This required some IT support and this capability varied at each site.

Neuromuscular Consent Form

This consists of informed consent about the participation in the database study for the patients with a neuromuscular disorder. At KUMC, the HSC-approved consent form is signed by the patient at the initial visit (Appendix 4). Other universities exempt the consent form in the initial visit and go to their HSC (IRB) for specific projects.

Codes for Muscular/Neuromuscular Junction Disease/ Peripheral Nervous System Disorders

Patients are assigned a diagnostic code at the initial visit. The database fields are minimal: patient's name, date of birth, and diagnostic code. These are entered into an Excel spreadsheet by a clinic clerk. If on the next visit there is a change of working diagnosis, the new code is entered into the computer spreadsheet.

At KUMC, the HSC-approved consent form is signed by the patient at the initial visit. Our goal is for all new clinic patients to sign the consent. This form states, "The database may be used to identify patients for research studies."

Results

Over 10 years, 17,163 of patients have been entered (Table 1). Table 2 shows the number of patients in several broad categories. In the motor neuron disease category, 2,023 of the 3,080 patients had amyotrophic lateral sclerosis (ALS) (Table 3). Of the 1,575 patients in the neuromuscular junction category, 1,368 had myasthenia gravis (Table 2). Of the 1,851 patients with muscular dystrophy, 258 had Duchenne muscular dystrophy and 330 had myotonic dystrophy (Table 3). Of the 633 inflammatory myopathy patients, 102 had dermatomyositis; 200 had inclusion body myositis; and 188 had polymyositis. Within the PNS group of patients with peripheral neuropathy, 1,090 had hereditary neuropathies; 1,001 had immune-mediated polyneuropathy; 620 had metabolic/toxic polyneuropathy; 535 had mononeuropathies; 296 had plexopathies; and 769 had radiculopathies. Table 4 shows the sum of related muscle disorders with the number of cases seen in this reporting period.

Discussion

Categorizing neuromuscular patients in tertiary care clinics is challenging. Although the ICD-10 numeric system is available, this is too crude and not sufficiently discriminating. For instance, both Myotonic dystrophy type 1 and type 2 have the same codes in the ICD-10 system.

This neuromuscular database is useful for determining the types of patients referred to tertiary care academic centers and more specific differentiation of the neuromuscular disorders. In the ICD-10 system, PLS, lower motor neuron disease and focal motor neuron disease have the same G12.29 codes whereas these have different codes in our neuromuscular database. Our database shows that in our tertiary care clinics, ALS is the most common occurring motor neuron disease. ALS was seen 13 times more than PLS. The database also reflects regional referral patterns. For example, because UTSW has a leprosy referral clinic, this diagnosis was unique to the Dallas clinic. Therefore, at UT San Antonio, these patients and diabetic patients tend to be seen in the general neurology clinic, which accounts for fewer patients in these categories in

this database.

The most common neuropathy seen in all four centers surprisingly was cryptogenic sensory polyneuropathy. This can be attributed to the tertiary care referral pattern. Within the diabetic neuropathy category, diabetic distal sensory polyneuropathy is seen three to four times more often than diabetic lumbosacral radiculoplexus neuropathy. This diagnostic frequency of diabetic polyneuropathy is still more common than would occur in a general medicine or neurology clinic and again indicates the tertiary care referral pattern. This also accounts for the many chronic inflammatory demyelinating polyneuropathy and multifocal motor neuropathy cases in the immune-mediated PNS group.

As expected, within the neuromuscular junction disorders, the most common neuromuscular junction disease was myasthenia gravis. MG is about 50 times more common than the Lambert-Eaton syndrome in our centers. The most common inflammatory myopathy disease seen was inclusion body myositis. Surprisingly, polymyositis cases were seen more often than dermatomyositis.[1,2]

Benefits of the neuromuscular database are:

1. Comparison of data between different hospitals.
2. Comparison between the initial visit diagnosis and final working diagnosis.
3. This intuitive and instinctive database is user friendly.
4. Allows continuous addition of new entities as they are discovered and reported.
5. Allows more precise coding of neuromuscular disorders beyond what can be done using the ICD system.

Finding an easy solution to enter the codes in the electronic medical record remained a barrier.

In summary, our experience shows that a diagnostic neuromuscular database can be utilized at multiple academic tertiary care centers. The database should be simple without too many fields to ensure compliance. This database has been very practical in identifying groups of patients for retrospective, observational studies and for prospective treatment studies (see tables 1-5).

Table 1. Total number of patients coded in four different hospitals of United States

Hospitals	Number of patients
UTHSCSA (1995-2005)	1,752
OSU (1995-2005)	1,840
KUMC (2001-2005)	3,699
UTSW (1993-2005)	9,872
TOTAL PATIENTS	17,163

Table 2. Total number of cases in different major categories seen in four hospitals.

Groups	UTSW	KUMC	OSU	UTHSCSA	Total
MOTOR NEURON DISEASE (MUS 100S)	1734	582	263	501	3080
NEUROMUSCULAR JUNCTION (MUS 200s)	1020	248	102	205	1575
MUSCULAR DYSTROPHIES (MUS 400s)	893	386	176	396	1851
HEREDITARY NEUROPATHIES (PNS 100s)	591	223	142	134	1090
IMMUNE MEDIATED POLYNEUROPATHY (PNS 200s)	285	225	257	234	1001
RADICULOPATHIES (PNS 700s)	542	206	6	15	769
INFLAMMATORY MYOPATHIES (MUS 500s)	375	88	96	74	633
METABOLIC/TOXIC POLYNEUROPATHY (PNS 300s)	368	131	112	9	620
MONONEUROPATHIES (PNS 500s)	221	209	48	57	535
PLEXOPATHIES (PNS 600s)	178	59	54	5	296

Table 3. Total number of cases in neuronopathies and neuropathies category seen in four hospitals.

DISEASE	UTSW	KUMC	OSU	UTHSCSA	TOTAL
ALS (MUS 101)	1051	414	147	411	2023
PLS (MUS 104)	86	31	12	19	148
CRYPTOGENIC SENSORY POLYNEUROPATHY (PNS 409)	191	210	270	24	695
DIABETES MELLITUS NEUROPATHY DSPN (PNS 309)	168	83	61	8	320
DIABETIC LUMBOSACRAL PLEXOPATHY (PNS 625)	41	29	20	0	90
CIDP (PNS 206)	177	72	61	32	342
MMN (PNS 209)	59	23	8	2	92

Table 4. Total number of cases in neuromuscular and myopathic category seen in four hospitals.

DISEASE	UTSW	KUMC	OSU	UTHSCSA	TOTAL
MYASTHENIA GRAVIS (MUS 201)	883	198	92	195	1368
LAMBERT EATON MYSTHENIC SYNDROME (MUS 202)	19	4	3	4	30
INCLUSION BODY MYOSITIS (MUS 504)	119	36	21	24	200
POLYMYOSITIS (MUS 501)	116	27	27	18	188
DERMATOMYOSITIS (MUS 502)	56	5	20	21	102
DUCHENNE MD (MUS 401)	42	53	35	128	258
BECKER MD (MUS 402)	35	11	2	26	74
FSH MD (MUS 405)	78	49	29	27	183
LGMD (MUS 404)	92	58	29	42	221
MYOTONIC DYSTROPHY (MUS421)	163	78	16	73	330
MYOTONIA CONGENITA (THOMSEN'S) (MUS 422)	26	8	4	13	51

REFERENCES

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2. Van der Meulen MGF, Bronner IM, Hoogendijk JE, Burger H, Van Venrooij WJ, Voskuyl AE, et al. Polymyositis: an overdiagnosed entity. *Neurology* 2003;61:316-21.

Appendix 1: Current MUS Codes

Category	Code		ICD-9	ICD-10
MUS	100	Motor Neuron Disorder	335.29	G12.29
MUS	101	Amyotrophic Lateral Sclerosis	335.2	G12.21
MUS	102	Familial ALS	335.2	G12.21
MUS	103	Lower Motor Neuron-MND	335.29	G12.29
MUS	104	Primary Lateral Sclerosis	335.24	G12.29
MUS	105	Familial Spastic Paraplegia	335.29	G11.4
MUS	106	Focal Motor Neuron Disease	335.29	G12.29
MUS	107	Stiff-Man Syndrome	333.91	G25.82
MUS	108	Isaac's Syndrome (Neuromyotonia NMT)	359.3	G71.19
MUS	109	Sequelae of Poliomyelitis	45.9	A80.9
MUS	110	Post-Polio Syndrome	138	G14
MUS	111	Benign Fasciculations	781	R25.3
MUS	112	ALS with frontotemporal dementia	335.2	G31.09
MUS	113	Other MND With frontotemporal dementia	335.29	G12.29/G31.09
MUS	120	Spinal Muscular Atrophy	335.1	G12.9
MUS	121	Infantile Onset SMA	335	G12.0
MUS	122	Childhood Onset SMA	335.11	G12.1
MUS	123	Adolescent Onset SMA	335.11	G12.1
MUS	124	Adult Onset SMA	335.19	G12.8
MUS	125	Distal SMA	335.1	G12.1
MUS	126	X-Linked SMA | Bulbospinal | Kennedy Syndrome	335.1	G12.2
MUS	130	Other Motor Neuron Disorder	335.29	G12.29
MUS	131	West Nile Virus Neuronopathy	335.29	A92.32
MUS	133	Hirayama’s Disease [Monomelic Amyotrophy]	335.29	G12.8
MUS	135	Hopkins Syndrome	335.29	Q87.0
MUS	136	IBALS [Isolated Bulbar ALS, MND]	335.22	G12.21
MUS	137	BAD [Brachial Amyotrophic Diplegia, MND]		
MUS	138	LAD [Leg Amyotrophic Diplegia, MND]		G82.20
MUS	139	Radiation-induced motor neuron disease	335.2	G12.29
MUS	200	Neuromuscular Junction Disorder	358.8	G70.89
MUS	201	Myasthenia Gravis [AchR Positive] (see 210 series)	358	G70.2
MUS	202	Lambert-Eaton Syndrome in neoplastic disease	358.1	G73.1

MUS	202	Lambert-Eaton Syndrome, unspecified	358.30	G70.80
MUS	202	Malignant Neoplasm		C80.1
MUS	203	Myasthenia Gravis [MuSK Positive]		
MUS	204	Magnesium Induced MG	358.2	G70.1
MUS	205	Antibiotic Induced MG	358.2	G70.1
MUS	206	Organophosphate Induced MG	358.2	G70.1
MUS	207	Organophosphate Induced Neuropathy	989.5	T63.89XA
MUS	208	Tick Paralysis	358.8	T48.1X#
MUS	209	Congenital Myasthenia Gravis	358	G70.2
MUS	210	MG, Ocular	358	G70.01
MUS	211	MG Status Post Thymectomy	358	G70.00
MUS	212	MG with Thymoma	358	G70.00
MUS	213	MG, in Remission	358	G70.00
MUS	214	MG, Asthenic bulbar	358	G70.00
MUS	215	MG, Crisis	358.01	G70.01
MUS	216	MG, Seronegative	358	G70.00
MUS	217	MG, Distal	358	G70.00
MUS	300	Muscle Disorder	359.9	G72.9
MUS	301	Myalgia	729.1	M79.1
MUS	301	Fibromyalgia	729.1	M79.7
MUS	302	Malignant Hyperthermia	995.86	T88.3
MUS	303	Rhabdomyolysis	728.88	M62.82
MUS	303	Myoglobinuria	791.3	R82.1
MUS	304	Fasciitis	729.4	M72.9
MUS	305	Hyper-CK-emia	790.5	R74.9
MUS	400	Muscular Dystrophy (MD)	359.1	G71.0
MUS	401	Duchenne MD	359.1	G71.0
MUS	402	Becker MD	359.1	G71.0
MUS	403	Other Dystrophinopathies	359.1	G71.0
MUS	404	Limb-Girdle MD (see 490 series)	359.1	G71.0
MUS	405	Facioscapulohumeral MD	359.1	G71.0
MUS	406	Scapuloperoneal MD	359.1	G71.0
MUS	407	Emery-Dreifuss MD	359.1	G71.0

MUS	408	Oculopharyngeal MD	359.1	G71.0
MUS	409	Distal MD	359.1	G71.0
MUS	410	Ocular Myopathy MD	359.1	G71.0
MUS	411	Other Muscular Dystrophy	359	
MUS	412	Muscular Dystrophy, Congenital	359	G71.2
MUS	413	Bethlem Myopathy	359	G71.0
MUS	414	Tubular Aggregate Myopathy	359	G71.2
MUS	415	Ulrich's Muscular Dystrophy	359.2	G71.0
MUS	420	Myotonic Disorders	359.2	G71.19
MUS	421	Myotonic Dystrophy, Type 1	359.2	G71.11
MUS	422	Myotonia Congenita, aut dominant	359.2	G71.12
MUS	423	Myotonia Congenita, aut recessive	359.2	G71.12
MUS	424	Paramyotonia Congenita	359.2	G71.19
MUS	425	Schwartz-Jampel (myotonic chondrodystrophy)	359.2	G71.13
MUS	426	Myotonic Dystrophy, Type 2	359.2	G71.11
MUS	427	Myotonic Dystrophy, unspecified	359	G72.9
MUS	430	Congenital Myopathy	359	G72.9
MUS	431	Centronuclear	359	G71.2
MUS	432	Nemaline Myopathy	359	G71.2
MUS	433	Central Core Myopathy		G71.2
MUS	434	Congenital Fiber Type Disproportion	359	G71.2
MUS	435	Hyaline Body Myopathy	359	G71.2
MUS	436	Congenital Hypotonia	359	P94.2
MUS	437	Reducing Body Myopathy	359	G71.2
MUS	438	Multi-Core	359	G71.2
MUS	439	Desmin Myopathy (SEE MUS 453 & 514)	359	G71.0
MUS	460	Metabolic Myopathy	359.8	E88.9
MUS	461	McArdle's Disease	271	E74.04
MUS	462	Other Glycogenoses	271	E74.09
MUS	463	Myoadenylate Deaminase Deficiency	359.8	E79.2
MUS	464	Muscle Carnitine Palmitoyltransferase Deficiency	272.9	E71.314
MUS	465	Other Lipid Disorders	272.9	E78.9
MUS	466	Kearns-Sayre Syndrome	359.8	H49.819
MUS	467	Other Mitochondrial Disorders	359.8	E88.49
MUS	468	Acid Maltase Deficiency [Pompe Disease]	359.8	E74.02

MUS	469	Danon Disease	359.8	E74.0
MUS	470	XEMA (X-Linked myopathy with excessive autophagy)	359	
MUS	471	CPEO (Chronic progressive external ophthalmoplegia)	378.72	H49.40
MUS	480	Periodic Paralysis (PP)	359.3	G72.3
MUS	481	Hypokalemic PP	359.3	G72.3
MUS	482	Hyperkalemic PP	359.3	G72.3
MUS	483	Normokalemic PP	359.3	G72.3
MUS	484	Andersen Tawil Syndrome	359.3	G72.3
MUS	485	Thyrotoxic PP	359.3	G72.3
MUS	486	LGMD, Type 1a	359.1	G71.0
MUS	487	LGMD, Type 1b	359.1	G71.0
MUS	488	LGMD, Type 1c	359.1	G71.0
MUS	489	LGMD, Type 1d	359.1	G71.0
MUS	490	LGMD, Type 2a	359.1	G71.0
MUS	491	LGMD, Type 2b / Miyoshi	359.1	G71.0
MUS	492	LGMD, Type 2c	359.1	G71.0
MUS	493	LGMD, Type 2d	359.1	G71.0
MUS	494	LGMD, Type 2e	359.1	G71.0
MUS	495	LGMD, Type 2f	359.1	G71.0
MUS	496	LGMD, Type 2g/ Distal myopathy / Telethonin	359.1	G71.0
MUS	497	LGMD, Type 2h	359.1	G71.0
MUS	498	LGMD, Type 2i	359.1	G71.0
MUS	499	LGMD, Type 2j	359.1	G71.0
MUS	500	Inflammatory Myopathies	710.4	G72.49
MUS	501	Polymyositis (organ involvement unspecified)	710.4	M33.20
MUS	502	Dermatomyositis	710.3	M33.90
MUS	503	Polymyositis with Connective Tissue Disease	710.4	M33.22
MUS	504	Inclusion Body Myositis (IBM)	359.71	G72.41
MUS	505	HIV Polymyositis	728	B20
MUS	506	Polymyalgia Rheumatica	725	M35.3
MUS	507	Sarcoid Myopathy (myositis)	135	D86.87
MUS	508	Viral Myositis	728	B97.89
MUS	509	Other Infectious Myopathies	728	M60.009
MUS	510	Eosinophilic Polymyositis	710.4	M33.20
MUS	511	Focal Myositis	729.1	M60.9

MUS	512	Necrotizing Myopathy	359.8	G72.89
MUS	513	Granulomatous Myositis	728.82	M60.20
MUS	550	LGMD 1F / TNPO3	359.1	G71.0
MUS	551	LGMD 1G / HNRPDL	359.1	G71.0
MUS	552	LGMD 1H / 3p23	359.1	G71.0
MUS	553	2K / MDDGC1 / POMT1	359.1	G71.0
MUS	554	2L / ANO5	359.1	G71.0
MUS	555	2M /MDDGC4 / Fukutin	359.1	G71.0
MUS	556	2N / MDDGC2 / POMT2	359.1	G71.0
MUS	557	2O / MDDGC3 / POMGnT1	359.1	G71.0
MUS	558	2P /MDDGC9 / DAG1	359.1	G71.0
MUS	559	2Q / Plectin 1f	359.1	G71.0
MUS	560	2R / Desmin	359.1	G71.0
MUS	561	2S / TRAPPC11	359.1	G71.0
MUS	570	HIBM1 / MFM1 / LGMD 1D / Desmin	359.71	G72.41
MUS	571	HIBM2 / Nonaka / GNE	359.71	G72.41
MUS	572	HIBM3 / Myosin heavy chain IIa	359.71	G72.41
MUS	573	HIBM with Paget disease & Dementia / VCP	359.71	G72.41
MUS	574	MFM2 / αB-crystalline	359	G72.89
MUS	575	MFM3 / LGMD 1A / Myotilin	359	G72.89
MUS	576	MFM4 / Markesbery-Griggs / ZASP	359	G72.89
MUS	577	MFM5 / Filamin C	359	G72.89
MUS	578	MFM6 / BAG3	359	G72.89
MUS	579	Myopathy & Respiratory Δ Type 1 / Udd / Titin	359	G72.89
MUS	580	Myopathy & Respiratory Δ Type 2 / 2q21	359	G72.89
MUS	581	Scapuloperoneal MD / Hyaline body myopathy / FHL-1	728.9	G71.0
MUS	582	Polyglucosan body / RBCK1	271	E74.09
MUS	583	Welander / TIA1	359.1	G71.0
MUS	584	Gowers-Laing / MPD1 / MYH7		G71.0
MUS	585	CMD with Desmin inclusions / SEPN1	359.1	G71.0
MUS	586	Distal Myopathy w/ vocal cord & Pharyngeal paralysis/ MPD2/ Matrin 3	359	G71.0
MUS	587	Miyoshi-like muscular dystrophy 2 / ? 10p	359.1	G71.0

MUS	588	Miyoshi-like muscular dystrophy 3 / Anoctamin 5 / ANO5	359.1	G71.0
MUS	589	Distal Nebulin myopathy / NEM2 Rod myopathy / Nebulin	359	G71.0
MUS	600	Other Myopathies	359.8	G72.89
MUS	601	Diabetic Muscle Infarction	359.8	
MUS	602	Toxic Myopathy	359.4	G72.2
MUS	603	Hyper Thyroid Myopathy		E05.90
MUS	603	Hypo Thyroid Myopathy		E03.90
MUS	604	Other Endocrine Myopathies	259.9	E34.9
MUS	605	Steroid Myopathy	359.4	G72.2
MUS	606	Amyloid Myopathy	277.3	E85.8
MUS	607	Muscle Tumors	239.3	C49.9
MUS	608	Alcoholic Myopathy	359.4	G72.2 (T51)
MUS	609	Thyroid Eye Disease [Graves' Eye Disease]	259.4	E05.00
MUS	610	Cholesterol Lowering Agent Myopathy [CLAM]	359.4	T46.6X#
MUS	611	Critical Illness Myopathy	359.8	G72.81
MUS	612	Isolated Neck Extensor Myopathy [INEM]	359.8	
MUS	613	[Statin-Associated] Autoimmune Necrotizing Myopathy		G72.81
MUS	700	EMG Abnormalities		
MUS	701	Martin-Gruber Anastomosis		
MUS	702	Accessory Peroneal Nerve		S84.10
MUS	703	Other Anomalous Innervation		
MUS	704	Conduction Block		R94.131
MUS	705	Pseudo-Conduction Block		
MUS	710	Abnormal Spontaneous Activity, Unspecified		R94.131
MUS	711	Cramps [General]	729.82	R25.2
MUS	712	Doublets/Multiplets		R94.131
MUS	713	Facial Myokymia		G51.4
MUS	714	Complex Repetitive Discharge [CRD]		R94.131
MUS	715	Neurotonia [on EMG]		G11.8
MUS	716	Myotonic Discharges		R94.131
MUS	717	Iterative Discharges		R94.131
MUS	718	Abnormal Insertional Activity		R94.131
MUS	719	Fasciculations	781	R25.3

MUS	720	Axon Reflex		R94.131
MUS	721	Stimulus Induced Repetitive Discharges		R94.131
MUS	722	Cramps Fasciculation Syndrome		R25.3
MUS	800	Limb-Girdle Syndrome	728.9	G71.0
MUS	801	Dropped Head Syndrome	780.79	M62.81
MUS	802	Dropped Body Syndrome	780.79	M62.81
MUS	803	Ptosis	378.9	H02.40
MUS	804	Ophthalmoplegia	378.9	H51.9
MUS	805	Dysarthria	784.5	R47.1
MUS	806	Dysphagia	784.5	R13.10
MUS	807	Ataxia	781.2	R27.0
MUS	808	Diplopia	368.2	H53.2
MUS	809	Bulbar Palsy	352.5	G12.22
MUS	810	Pseudobulbar Palsy	335.23	G12.8
MUS	811	Fatigue, NOS	780.79	R53.83
MUS	812	Upper Extremity Weakness	728.9	G83.20
MUS	813	Lower Extremity Weakness	728.9	G83.10
MUS	814	Hand Weakness	728.9	M62.81
MUS	815	Neck Weakness	728.9	M62.81
MUS	816	[Neuromuscular] Respiratory Weakness	780.79	R53.83
MUS	817	Exercise-Induced Weakness	780.79	R53.83
MUS	818	PEO [Progressive External Ophthalmoplegia]		H49.4
MUS	900	Neuromuscular Disorders		G70.9
MUS	901	Chronic Fatigue Syndrome	780.71	R53.82
MUS	910	Related to Cancer	239.9	D49.9
MUS	911	Thymoma [Benign]	212.6	D15.0
MUS	911	Thymoma [Malignant]	164.0	C37
MUS	920	Hereditary		
MUS	921	Dominant		
MUS	922	Recessive		
MUS	923	X-Linked		
MUS	930	Myelopathy	336.9	G95.9
MUS	931	B12 Deficiency	266.9/281.1	E53.8
MUS	932	Vitamin E Deficiency	269.1	E56.0
MUS	933	HIV Related	42	B20

MUS	934	HTLV-1 Related [Myelopathy]	42	C91.50
MUS	935	Cervical Myelopathy	336.9	M47.12
MUS	936	Thoracic Myelopathy	336.9	M47.14
MUS	937	Lumbosacral Myelopathy	336.9	M47.16
MUS	938	Cauda-Equina Syndrome	344.6	G83.4
MUS	939	Syringomyelia & syringobulbia	336	G95.0
MUS	940	Degenerative Spine Diseases [Spondylosis]	722.6	M51.9
MUS	941	Meningomyelocele [Spina Bifida]	741.9	Q05.9
MUS	942	Spinocerebellar Degeneration	334.9	G11.9
MUS	943	Traumatic [Injury]	959.9	T14.90
MUS	944	Spasticity, NOS	781.2	R25.2

Appendix 2: Current PNS Codes

Category	Code		ICD-9	ICD-10
PNS	100	Inherited Polyneuropathy	356	G60.0
PNS	101	CMT Type 1, (see PNS 130 series)	356.1	G60.0
PNS	102	CMT Type 2, (see PNS 140 series)	356.1	G60.0
PNS	103	CMT Type 3	356.1	G60.0
PNS	104	X-Linked CMT	356.1	G60.0
PNS	105	Familial Amyloidosis	277.3	E85.9
PNS	106	Porphyria	277.1	E80.2
PNS	107	Leukodystrophy	330	E75.29
PNS	108	Refsum disease	356.3	G60.1
PNS	109	Glycogen Storage Disease	330.9 (271.0)	E74.00
PNS	110	Hereditary Neuropathy with Pressure Palsies [HNPP]	356.9	G60.0
PNS	111	Friedreich's Ataxia	334	G11.1
PNS	112	Neuropathy in Olivopontocerebellar Atrophy [OPCA]	333	G62.9 / G23.8
PNS	113	Neuropathy in Other Multiple System Degeneration	337.9	G62.9 / G90.3
PNS	115	Neuropathy in Neurofibromatosis		G62.9 / Q85.00
PNS	116	Polyglucosan Body Disease		E74.09
PNS	117	Fabry Disease	272.7	E75.21
PNS	120	Hereditary sensory/autonomic neuropathy (HSN/HSAN)	356	G60.9
PNS	130	CMT 1A	356.1	G60.0
PNS	131	CMT 1B	356.1	G60.0
PNS	132	CMT 1C	356.1	G60.0
PNS	133	CMT 1D	356.1	G60.0
PNS	134	CMT 1E	356.1	G60.0
PNS	135	CMT 1F	356.1	G60.0
PNS	140	CMT 2A	356.1	G60.0
PNS	141	CMT 2B	356.1	G60.0
PNS	142	CMT 2C	356.1	G60.0
PNS	143	CMT 2D	356.1	G60.0
PNS	144	CMT 2E	356.1	G60.0
PNS	145	CMT 2F	356.1	G60.0
PNS	146	CMT 2G	356.1	G60.0
PNS	147	CMT 2H	356.1	G60.0
PNS	148	CMT 2I	356.1	G60.0
PNS	149	CMT 2J	356.1	G60.0

PNS	150	CMT 2I	356.1	G60.0
PNS	160	CMT Type 4, (autosomal recessive)	356.1	G60.0
PNS	170	HMSN 5	356.1	G60.0
PNS	180	HMSN 6	356.1	G60.0
PNS	181	Hereditary Neuralgic Amyotrophy	356.1	G54.5
PNS	182	Familial Amyloid Neuropathy	277.3	E85.1
PNS	190	HMN	335.29	G60.0
PNS	200	Immune Mediated/Inflammatory/Infectious Polyneuropathy	357.8	G61.89
PNS	201	Guillain-Barre Syndrome	357	G61.0
PNS	202	Recurrent GBS	357	G61.0
PNS	203	Miller-Fisher Syndrome	357	G61.0
PNS	204	Axonal GBS	357	G61.0
PNS	205	Acute Paralytic Poliomyelitis	357	A80.30
PNS	206	Chronic Inflammatory Demyelinating Polyneuropathy	357.81	G61.81
PNS	207	Neuropathy Due to Vasculitis	357.1	G63
PNS	208	Neuropathy Due to other Connective Tissue Disease (see 212 series)	357.1	G63
PNS	209	Multifocal Motor Neuropathy	357.9	G60.9
PNS	210	CIDP with CNS Overlap	357.8	G61.81
PNS	211	MADSAM neuropathy (Multifocal Acquired Demyelinating Sensory and Motor neuropathy)	357.81	G61.8
PNS	212	Neuropathy Due to SLE	357.1	M32.19
PNS	213	Neuropathy Due to RA	357.1	M05.50
PNS	214	Neuropathy Due to Sjogren's Syndrome	357.1	M35.00
PNS	215	Neuropathy Due to Peripheral Vascular Disease	357.4	G63
PNS	220	Neuropathy Due to Infection	357.9	G61.9
PNS	221	Neuropathy Due to Lyme Disease	357.9	A69.22
PNS	222	Neuropathy Due to Herpes Zoster	357.9	B02.23
PNS	223	Neuropathy Due to HIV	357.9	B20
PNS	224	Neuropathy Due to Leprosy	357.9	A30.9
PNS	225	Sarcoid neuropathy		D86.87
PNS	239	DADS neuropathy (Distal Acquired Demyelinating Symmetric neuropathy)		
PNS	240	Neuropathy Associated with Monoclonal Protein	357.9	G61.9
PNS	241	Neuropathy Associated with MGUS	357.9	G61.9
PNS	242	Neuropathy Associated with POEMS	357.9	G61.9
PNS	243	Neuropathy Associated with Anti-MAG	357.9	G61.9
PNS	244	Neuro. Assoc. w/ Monoclonal Protein Due to Amyloidosis	277.3	E85.1, 3, 8

PNS	245	Neuropathy Associated with GM-1 Antibody	357.9	G61.9
PNS	246	Neuropathy Associated with Multiple Myeloma	357	C90.00
PNS	247	Neuropathy Associated with Anti-Hu Antibody	357.9	G61.9
PNS	248	Paraneoplastic Neuropathy [non-Hu]	357.9	G61.9
PNS	249	Anti-AchR Antibody Neuropathy		
PNS	250	Multiple Sclerosis	340	G35
PNS	251	Neuropathy Due to Hepatitis B	357.4	G63
PNS	252	Neuropathy Due to Hepatitis C	357.4	G63
PNS	255	Neuropathy Due to Prior GVHD	357.4	G63
PNS	260	Neuropathy Due to Celiac Disease	357.4	G63
PNS	261	Neuropathy Due to Inflammatory Bowel Disease	357.4	G63
PNS	262	CANOMAD syndrome	357.81	G61.8
PNS	270	Neuropathy Due to Myelodysplasia	357.4	G63
PNS	271	Potassium channel antibody syndrome		
PNS	300	Metabolic Neuropathy	357.9	G61.9
PNS	301	Renal Disease Neuropathy	357.9	G61.9
PNS	302	Hepatic Disease Neuropathy	357.9	G61.9
PNS	303	Thyroid Disease Neuropathy	357.9	G61.9
PNS	304	Pregnancy Neuropathy	357.9	G61.9
PNS	305	Critical-Illness Neuropathy	357.9	G61.9
PNS	306	Nutritional Neuropathy	263.8	E46
PNS	307	Vitamin B12 Deficiency Neuropathy	266.2/281.1	E53.8
PNS	308	Vitamin E Deficiency Neuropathy	269.1	E56.9
PNS	309	Diabetes Mellitus Sensorimotor Polyneuropathy	357.2/250.6	E11.40
PNS	311	Diabetic Neuropathic Cachexia	250.9	E11.40
PNS	312	Impaired Glucose Tolerance Neuropathy		R73.02
PNS	320	Toxin Neuropathy	357.7	G62.2
PNS	321	Alcohol Neuropathy	357.5	G62.1
PNS	322	Heavy Metals Neuropathy	357.7	G62.2
PNS	323	Copper/Zinc-related Myeloneuropathy	357.4	
PNS	330	Drug Neuropathy	357.6	G62.0
PNS	331	Chemotherapeutic Drug Neuropathy	357.6	G62.0
PNS	332	Dilantin Neuropathy	357.6	G62.0
PNS	333	Hypertriglyceridemia Neuropathy		E78.1

PNS	400	Polyneuropathy, NOS	357.9/356.4	G62.3
PNS	401	Sensory Neuropathy	357.9/356.4	G60.8
PNS	402	Acute Motor Neuropathy	357.9/356.4	G62.81
PNS	403	Sensorimotor Neuropathy	357.9/356.4	G60.8
PNS	404	Autonomic Neuropathy	337	G90.9
PNS	405	Axonal Neuropathy	357.9/356.4	G61.9
PNS	406	Demyelinating Neuropathy	357.9/356.4	G61.9
PNS	407	Mixed Axonal Demyelinating Neuropathy	357.9/356.4	G61.9
PNS	408	Sensory Neuronopathy (Ganglionopathy) - autoimmune autonomic ganglionopathy	357	G90.0
PNS	409	Cryptogenic Sensory Polyneuropathy (CSPN)	357.9/356.4	G61.9
PNS	410	Small-Fiber Neuropathy	357.9/356.4	G61.9
PNS	411	DADS Neuropathy (Distal acquired demyelinating)	357.81	G61.81
PNS	412	MAMA (Multifocal acquired motor axonal)	357.81	G61.81
PNS	413	POTC		
PNS	500	Mononeuropathy	355.9	G58.9
PNS	501	Radial Neuropathy	354.3	G56.3
PNS	502	Posterior Interosseous Neuropathy	354.3	G56.3
PNS	503	Axillary Neuropathy	353	G56.90
PNS	510	Median Neuropathy	354.1	G56.1
PNS	511	Carpal Tunnel Syndrome	354	G56.0
PNS	512	Median Neuropathy in the Arm	354.1	G56.10
PNS	513	Anterior Interosseus Neuropathy	354.8	G56.80
PNS	520	Ulnar Neuropathy	354.2	G56.20
PNS	521	Ulnar Neuropathy at the Wrist	354.2	G56.20
PNS	522	Ulnar Neuropathy at the Ulnar Grove	354.2	G56.20
PNS	530	Peroneal Neuropathy	355.36	G57.30
PNS	531	Peroneal Neuropathy at the Knee	355.36	G57.30
PNS	540	Facial Neuropathy	351.9	G51.9
PNS	541	Bell's Palsy	351	G51.0
PNS	542	Facial Neuropathy-Traumatic	351.9	G51.9
PNS	543	Facial Neuropathy Due to Surgery	351.9	G51.9
PNS	544	Facial Neuropathy Due to Infection	351.9	G51.9
PNS	545	Pain due to Neuropathy of Facial Nerve	351.8	G51.8
PNS	546	Hemifacial Spasm	351.8	G51.3
PNS	550	Trigeminal Neuropathy	350.1	G50.0

PNS	551	Trigeminal Neuropathy-Herpes Zoster	53.1	B02.29
PNS	552	[Spinal] Accessory Neuropathy	352.4	G52.8
PNS	553	Hypoglossal Neuropathy	352.5	G52.3
PNS	554	Cranial Neuropathy	352.9	G52.9
PNS	560	Musculocutaneous Neuropathy	354.8	G56.80
PNS	561	Suprascapular Neuropathy	354.8	G56.80
PNS	562	Long Thoracic Neuropathy	354.8	G56.80
PNS	563	Upper Extremity Neuropathy	354.9	G56.90
PNS	570	Tibial Neuropathy	355.79	G56.80
PNS	571	Sciatic Neuropathy	355	G57.00
PNS	572	Femoral Neuropathy	355.2	G57.20
PNS	572	Femoral Neuropathy [R lower limb]		G75.21
PNS	572	Femoral Neuropathy [L lower limb]		G75.22
PNS	573	Saphenous Neuropathy	355.79	G57.80
PNS	574	Obturator Neuropathy	355.79	G57.80
PNS	575	Sural Neuropathy	355.79	G57.80
PNS	576	Plantar Neuropathy	355.79	G57.60
PNS	577	Other Lower Extremity Neuropathy	355.79	G57.90
PNS	578	Lateral Femoral Cutaneous Neuropathy	355.1	G57.10
PNS	579	Peroneal Neuropathy	355.9	G57.30
PNS	580	Phrenic Neuropathy	G58.8	
PNS	600	Celiac Plexopathy		G54.8
PNS	610	Brachial Plexus Neuropathy	353	G54.0
PNS	611	Idiopathic Brachial Plexus Neuropathy	353.8	G54.0
PNS	612	Traumatic Brachial Plexus Neuropathy	353.8	G54.0
PNS	613	Radiation Brachial Plexus Neuropathy	353.8	G54.0
PNS	614	Malignancy Brachial Plexus Neuropathy	353.8	G54.0
PNS	615	Diabetic Brachial Plexus Neuropathy	358.1	G54.0
PNS	616	Hematoma Brachial Plexus Neuropathy	353.8	G54.0
PNS	617	Hereditary Brachial Plexus Neuropathy		G54.0
PNS	620	Lumbosacral Plexus Neuropathy	353.1	G54.1
PNS	621	Idiopathic Lumbosacral Plexus Neuropathy	353.1	G54.1
PNS	622	Traumatic Lumbosacral Plexus Neuropathy	353.8	G54.1
PNS	623	Radiation Lumbosacral Plexus Neuropathy	353.8	G54.1
PNS	624	Malignancy Lumbosacral Plexus Neuropathy	353.8	G54.1
PNS	625	Diabetic Lumbosacral Plexus Neuropathy, (Amyotrophy)	358.1	G54.1

PNS	626	Hematoma Lumbosacral Plexus Neuropathy	353.8	G54.1
PNS	627	Hereditary Lumbosacral Plexus Neuropathy		G54.1
PNS	700	Radiculopathy	729.2	M54.10
PNS	710	Cervical Radiculopathy	723.4	M54.12
PNS	711	Cervical Radiculopathy - Disc	722	M50.10
PNS	712	Cervical Radiculopathy - Degenerative Spine Disease	722	M50.10
PNS	713	Cervical Radiculopathy - Diabetes	723.4	M54.12
PNS	714	Cervical Radiculopathy - Tumor	723.4	M54.12
PNS	715	Cervical Radiculopathy - Trauma	722	M54.12
PNS	716	Cervical Radiculopathy - Herpes Zoster	53.19	B02.29
PNS	720	C5 Cervical Radiculopathy	722	M50.12
PNS	721	C6 Cervical Radiculopathy	722	M50.12
PNS	722	C7 Cervical Radiculopathy	722	M50.12
PNS	723	C8 Cervical Radiculopathy	722	M50.13
PNS	730	Thoracic Radiculopathy	724.4	M54.14
PNS	731	Thoracic Radiculopathy - Disc	722.11	M51.14
PNS	732	Thoracic Radiculopathy - Degenerative Spine Disease	722.11	M51.14
PNS	733	Thoracic Radiculopathy - Diabetes	724.4	M54.14
PNS	734	Thoracic Radiculopathy - Tumor	724.4	M54.14
PNS	735	Thoracic Radiculopathy - Trauma	722.11	M51.34
PNS	736	Thoracic Radiculopathy - Herpes Zoster	53.19	B02.29
PNS	740	Lumbosacral Radiculopathy	724.4	M54.17
PNS	741	Lumbosacral Radiculopathy - Disc	722.1	M51.17
PNS	742	Lumbosacral Radiculopathy - Degenerative Spine	722.1	M51.17
PNS	743	Lumbosacral Radiculopathy - Diabetes	724.4	M54.17
PNS	744	Lumbosacral Radiculopathy - Tumor	724.4	M54.17
PNS	745	Lumbosacral Radiculopathy - Trauma	722.1	M51.27
PNS	746	Lumbosacral Radiculopathy - Herpes Zoster	53.19	B02.29
PNS	747	Lumbosacral Radiculopathy - Other	724.4	M54.17
PNS	748	Lumbosacral Radiculopathy - HIV	724.4	M54.17
PNS	750	Lumbosacral Radiculopathy - L1	722.1	M51.27
PNS	751	Lumbosacral Radiculopathy - L2	722.1	M51.27
PNS	753	Lumbosacral Radiculopathy - L3	722.1	M51.27
PNS	754	Lumbosacral Radiculopathy - L4	722.1	M51.27
PNS	755	Lumbosacral Radiculopathy - L5	722.1	M51.27

PNS	756	Lumbosacral Radiculopathy - S1	722.1	M51.27
PNS	757	Lumbosacral Spinal Stenosis	724.02	M48.07
PNS	758	Spinal AVM	G27.39	
PNS	800	Mononeuritis Multiplex	354.5	G58.7
PNS	801	Mononeuritis Multiplex - Vasculitis	354.8	G58.7
PNS	802	Mononeuritis Multiplex - Diabetes	354.8	G58.7
PNS	803	Mononeuritis Multiplex - HIV	354.8	G58.7
PNS	804	Mononeuritis Multiplex - CMV	354.8	G58.7
PNS	805	Mononeuritis Multiplex - Other Infectious	354.8	G58.7
PNS	806	Mononeuritis Multiplex - Collagen Vascular Disease	354.8	G58.7
PNS	807	Mononeuritis Multiplex - Tumor	354.8	G58.7
PNS	808	Mononeuritis Multiplex - HNPP	356.1	G58.7
PNS	820	Pain, NOS	729.1	R52.0
PNS	821	Hand Pain	729.5	M79.64
PNS	822	Shoulder Pain	719.41	M25.51
PNS	823	Upper Extremity Pain, NOS	729.5	M79.609
PNS	824	Foot Pain	729.5	M79.67
PNS	825	Knee Pain	719.46	M25.56
PNS	826	Hip Pain	719.45	M25.55

PNS	827	Lower Extremity Pain, NOS	729.5	M79.609
PNS	828	Neck Pain	729.1	M54.2
PNS	829	Back Pain	724.2	M54.9
PNS	830	Abdominal Pain	789	R10
PNS	831	Chest Pain	786.59	R07.9
PNS	832	Cephalgia	784	R51
PNS	833	Reflex Sympathetic Dystrophy (RSD)	337.2	G90.59
PNS	834	Painful Legs Moving Toes	729.5	M79.609
PNS	835	Numbness, NOS/Parasthesias	782	R20.2
PNS	836	Upper Extremity Numbness	782	R20.2
PNS	837	Lower Extremity Numbness	782	R20.2
PNS	838	Face Numbness	782	R20.2
PNS	839	Ataxia	781.2	R27.0
PNS	840	Conversion Disorder	300.11	F44.4
PNS	841	Somatic Complaints	300.81	F45.0
PNS	842	Erythromelalgia	443.82	I73.81
PNS	910	Neuropathy Associated with Cancer	199.1	C80.1
PNS	911	Lymphoma Related Neuropathy	202.8	
PNS	912	Small-Cell Tumor Related Neuropathy	199.1	C80.1
PNS	920	Hereditary		
PNS	921	Dominant		
PNS	922	Recessive		
PNS	923	X-Linked		
CNS	100	Movement Disorders	333.9	G25.9
CNS	110	Tremor	333.1	R25.1
CNS	120	Parkinsonism	332	G20.0
CNS	130	Dystonia	333.6	G24.9
CNS	140	Chorea	333.5	G25.5
CNS	141	Restless Legs Syndrome (RLS)	333.99	G25.81
CNS	200	Seizure Disorders	345.9	G40.909
CNS	300	Dementia	290	F03.90
CNS	301	Alzheimer Disease	331	G30.9
CNS	302	Frontotemporal dementia	290	G31.09
CNS	400	Gait Disturbance	781.2	R26.9

CNS	500	Cerebral Palsy	343.9	G80.9

Appendix 3: Data Collection Sheet

Neuromuscular Clinical Data Base

Patient Name: _____

KU ID #: _____

DOB: _____

SS #: _____

NM Code: _____

ICD-10 Code: _____

Date of Service: _____

Signature: _____

Date: _____

Appendix 4: KUMC IRB consent form

Please feel free to read but DO NOT SIGN until your doctor has discussed this with you.

Thank you

Neuromuscular Database Project Informed Consent

INTRODUCTION

I understand that I am being asked to participate in a database study for patients with a neuromuscular disorder. If I consent, the information collected during my clinic visits will be entered into a database. This information is gathered in the form of medical record number, name, date of birth, date of visit, and diagnosis code. An arbitrary neuromuscular code will be assigned based on my diagnosis. Dr. Mazen Dimachkie is conducting this study at the KUMC Center for Neuromuscular Disease. Every patient who presents in the neuromuscular clinic will be asked to participate.

You do not have to participate in this research study. It is important that before you make a decision to participate, read the rest of this form. You should ask as many questions as needed to understand what will happen to you if you participate in this study.

BACKGROUND

The Neuromuscular Clinic offers diagnostic and treatment services for persons with Neuromuscular disease and other related conditions. Patients may also participate in clinical research pertaining to investigational medications.

PURPOSE

The purpose of this project is to collect information on neuromuscular patients into a database and to use this information for scientific study and improvement of treatment procedures. This database may be used to identify patients for research studies.

If I am identified through this database for potential participation in a research study, I:

____ give my permission
____ do not give my permission

to be contacted regarding these studies. By signing this consent form, I am under no obligation to participate in any future research study. I would be contacted by the neuromuscular research nurse, solely under the direction of one of the doctors treating me in the neuromuscular clinic.

PROCEDURE

Should I decide to participate in this database study, information collected during my normal clinic visits will be entered into a computer database along with information from other patients from the Neuromuscular Clinic. Information obtained during routine clinic visits may be entered into a separate database for study purposes. This includes any testing performed by members of the clinic staff; i.e. speech therapist, social worker, and others that I see during the course of the clinic visit. Other information in my KU records related to my neuromuscular disease may be entered into the research database. Participating in this study will not add to the length of my normal clinic visit. If I consent, the

data will be collected during my initial visit and then at each follow-up visit.

Information in the Neuromuscular Database will be analyzed for research purposes. Researchers may at various times study topics such as aspects of my disease, effects of standard medications, and quality of life.

Researchers at KUMC will analyze information in the database and they may share it with researchers at other universities. If study data is shared outside KUMC, individual identifiers will be removed so that the participants' identities are not known.

RISKS

I am taking no foreseeable risks by participating in this study. The information I disclose will remain confidential.

NEW FINDINGS STATEMENT

I will be informed if any significant new findings develop during the course of the study that may affect my willingness to participate in this study.

BENEFITS
I will not directly benefit from participating in this study, however the information collected will be used for scientific study and may result in improved treatment of neuromuscular disorders.

PAYMENT TO THE SUBJECT
I will not receive payment for participating in this study. Also, I will retain my current financial responsibility for my visits to the Neuromuscular Clinic.

COSTS

There is no additional cost to me for participating in this study.

ALTERNATIVES

My alternative is to not participate in the Neuromuscular Database Project.

INSTITUTIONAL DISCLAIMER STATEMENT

If you think you have been harmed as a result of participating in research at the University of Kansas Medical Center (KUMC), you should contact the Director, Human Research Protection Program, Mail Stop #1032, University of Kansas Medical Center, 3901 Rainbow Blvd., Kansas City, KS 66160. Under certain conditions, Kansas state law or the Kansas Tort Claims Act may allow for payment to persons who are injured in research at KUMC.

CONFIDENTIALITY AND PRIVACY AUTHORIZATION

My name or information identifying me will not be released without my written permission unless required by law. Study data will be recorded on the Neuromuscular Database form and entered into the database by the Research Instructor. Researchers cannot guarantee absolute confidentiality. If any information obtained from this database is published or presented in public, information that identifies me will be removed.

The privacy of my health information is protected by a federal law known as the Health Insurance Portability and Accountability Act (HIPAA). If I choose to participate in this study, I will be asked to give permission for researchers to use and disclose my health information.

To perform this study, researchers will collect health information about me from my medical record and from the study activities that are listed in the Procedures section of the consent form. My study-related health information will be used at KU Medical Center by Dr. Mazen Dimachkie, members of the research team, the KU Hospital Medical Record

Department, the KUMC Research Institute and officials at KUMC that oversee research, including the KUMC Human Subjects Committee, and federal officials who oversee research, if a regulatory review takes place.

All study information that is sent outside KU Medical Center will have my name and other identifying characteristics removed, so that my identity will not be known. Because identifiers will be removed, my health information will not be re-disclosed by outside persons or groups and will not lose its federal privacy protection.

Permission granted on this date to use and disclose my health information remains in effect indefinitely. By signing this form I give permission for the use and disclosure of my information for purposes of the study at any time in the future. Any research information that is placed in my medical record will be kept indefinitely.

QUESTIONS

I have read the information in this form. The investigators have answered my questions to my satisfaction. I know if I have any more questions after signing this form, I may contact Mazen Dimachkie, MD at (913) 588-6970. If I have any questions about my rights as a research subject, I may call (913) 588-1240 or write the Human Subjects Committee at Mail Stop #1032, University of Kansas Medical Center, 3901 Rainbow Blvd., Kansas City, KS 66160.

SUBJECT RIGHTS AND WITHDRAWAL FROM THE STUDY

I understand that my participation in this study is voluntary and that the choice not to participate or to quit at any time can be made without penalty or loss of benefits. Deciding not to participate or quitting will have no effect upon the medical care or treatment I receive now or in the future at the University of Kansas Medical Center.

If I want to cancel permission to use your health information, I should send a written request to Dr. Mazen Dimachkie. The mailing address is Mazen Dimachkie, M.D., University of Kansas Medical Center, 3599 Rainbow Blvd, Mail Code 2012, Kansas City, KS 66160. If I cancel permission to use my health information, the research team will stop collecting any additional information about me. All of the data collected on me will be removed from the database if I request it.

CONSENT

The investigators gave me information about what will be done to me in this research study. The also told me how it will be done, what I will have to do, and how long the research will take. They told me about any inconvenience, discomfort or risks I might experience due to this research. They explained to me how this research might affect my health or me. I agree to take part in this study as a research subject. I am aware that I may quit or refuse any part of this research study at any time. I understand that quitting will have no effect upon this medical care or treatment I receive in the future.

By signing this form, I give my permission for my health information to be used and disclosed for the purposes of this research study. If I choose not to sign this form, my information will not be entered into the database. The research team may use and share information that was gathered before they received your cancellation.

I understand that the investigators will give me a signed copy of this form to keep for my records.