

**Myotonic Disorders of Muscle – Table 1**

	<b>Myotonic Dystrophy (DM1)</b>	<b>Myotonic Dystrophy (DM2)</b>	<b>Myotonia Congenita</b>	<b>Paramyotonia Congenita</b>	<b>Hypokalemic Periodic Paralysis</b>	<b>Hyperkalemic Periodic Paralysis</b>
	<i><b>Dystrophic (multisystem involvement)</b></i>		<i><b>Non-dystrophic (exclusive skeletal muscle dysfunction)</b></i>			
<b>Inheritance</b>	AD	AD	AD (Thomsen) AR (Becker)	AD	AD	AD
<b>Ion Channel Affected</b>	Chloride, Sodium	Chloride, Sodium	Chloride	Sodium	Calcium, Sodium	Sodium
<b>Gene</b>	DMPK	CNBP (ZNF9)	CLCN1	SCNA4	CACNL1A3or SCNA4	SCNA4
<b>Chromosome</b>	19q13	3q21	7q35	17q23-25	1q31-32, 17q23-25	17q23-25
<b>Myopathy</b>	Distal	Proximal	Rare	Very Rare	Common	Rare
<b>Triggers</b>	Exercise	Exercise	Sudden movement, cold	Cold, rest after exercise	Rest after exercise, high carbohydrates, sodium	Cold, fasting, exercise, potassium ingestion
<b>Therapy (avoid triggers)</b>			Mexiletine Exercise	Mexiletine	Potassium Acetazolamide (Diamox)	Thiazides Tocainide
<b>Classic features</b>			Improves with repetitive muscle activity (warm up phenomenon)	Worsens with repetitive muscle activity		