Table 1. Updated classifications of dystonia over last 20 years

	Fahn, Bressman, and Marsden (1998)	**Fahn, Jankovic, and Hallett (2011)**	**EFNS guidelines 2011**	**Consensus statement 2013[1] (Axis 1 clinical axis 2 etiology)**
Age at onset	1.Childhood 2.Adolescence 3.Adulthood	1.Young-onset <26 2. Adult-onset >26	1.Early-onset (variably defined as <20–30 years) 2.Late-onset	1. Infancy (birth to 2 years) 2. Childhood (3–12 years) 3. Adolescence (13–20 years) 4. Early adulthood (21–40 years) 5. Late adulthood (>40 years)
Distribution	Focal Segmental Multifocal Generalized Hemidystonia	Focal Segmental Multifocal Generalized Hemidystonia	Focal Segmental Multifocal Generalized Hemidystonia	Focal Segmental Multifocal Generalized (with or without leg involvement) Hemidystonia
Temporal pattern	N.A.	N.A.	N.A.	1. Disease course a. Static b. Progressive 2. Variability a. Persistent b. Action-specific c. Diurnal d. Paroxysmal
Associated Features				Isolated dystonia or combined with another movement disorder 1. Isolated dystonia 2. Combined dystonia Occurrence of other neurological or systemic manifestations 1. List of co-occurring neurological manifestations
Aetiology	**Primary** a. Familial b. Sporadic	**Primary**	**Primary** a. Primary pure (eg DYT1,6) b. Primary paroxysmal (eg DYT8,10,18)	**Axis II Aetiology** **Nervous system pathology** 1. Evidence of degeneration 2. Evidence of structural (often static) lesions 3. No evidence of degeneration or structural lesion **Inherited or aquired** **Inherited** 1. Autosomal dominant 2. Autosomal recessive 3. X-linked recessive
	Dystonia-plus	**Dystonia-plus**	c. Primary plus (former dystonia-plus syndromes eg DYT5,11)	
	Secondary	**Secondary**	**Secondary**	
	Heredo-degenerative	**Heredo-degenerative** **Feature of another**	**Heredo-degenerative**	

		neurologic disease (eg, dystonic tics,paroxysmal dyskinesias, PD, PSP etc)		4. Mitochondrial **Acquired** 1. Perinatal brain injury 2. Infection 3. Drug 4. Toxic 5. Vascular 6. Neoplastic 7. Brain injury *8. Psychogenic(see text* Idiopathic 1. Sporadic *2.* Familial*)*

Abbreviations: EFNS – European federation of neurological sciences **, N.A.** Not applicable

Table 2 Identification of aetiology for dystonia using the consensus statement 2013 [1]

Axis 1 Clinical characteristics				Axis 2 Etiology
Onset age	Isolated/ combined/ Specific features	Course/Variability	Focal/ generalized	Inherited/Acquired/ Structural/ Idiopathic
Childhood	Isolated	Progressive	Generalized	**Inherited AD** *DYT1* *DYT 6* DRD (DYT 5a)
Childhood to adulthood	Isolated	Progressive	Focal/ segmental/ generalized	**Inherited AD** *DYT6*
Childhood to adulthood	Isolated/ combined with pyramidal/ parkinsonism/ myoclonus	Static	Focal/ segmental/ generalized	**Nervous system pathology without degeneration** Perinatal injury Vascular Toxin Brain injury
Childhood	Combined with pyramidal/ parkinsonism/ myoclonus	Progressive	Focal/ segmental/ multifocal/generalized	**Nervous system pathology with degeneration** (or heredodegenerative) like Wilson's, NBIA, mitochondrial etc.
Adulthood/ Adolescence to adulthood	Isolated	Progressive	Focal/ segmental	**Inherited AD** *DYT 23* *DYT 24* *DYT25*
Childhood	Combined with Parkinsonism	Progressive but with diurnal fluctuation	Generalized	**Inherited AD** *DYT5a*
Adulthood	Combined with Parkinsonism	Progressive	Generalized	**Inherited XL** *DYT3*
Childhood-adulthood	Combined with Parkinsonism	Progressive	Focal/segmental/ generalized	**Inherited AD** *DYT16* RODP **Inherited AR** DTD YOPD **Acquired** **Inherited AD** (Huntington's disease,

				Spinocerebellar ataxia) **Acquired** **Inherited AR** GM1 gangliosidosis **Nervous system pathology with degeneration** MSA PD
Infancy-	Combined	Progressive	Generalized	**Inherited AR** *DYT5b*
Childhood/adulthood	Combined with myoclonus or parkinsonism	Progressive	Focal/Generalized	**Nervous system pathology with degeneration** (or heredodegenerative) Wilson's disease GM1 and GM2 gangliosidosis Glutaric acidemia type I Homocystinuria Lesch-Nyhan disease Leigh's disease Primary familial brain calcification Neurodegeneraton with brain iron accumulation Dystonic lipodosis Ceroid lipofusinosis Ataxia telengiectaisa Neuroacanthocytosis
Childhood-adolescence	Combined with myoclonus	Progressive	Segmental/ generalized	**Inherited AD** *DYT11* (myoclonus dystonia)
Childhood-adolescence	Isolated	Paroxysmal	Focal/ generalized	**Inherited AD** *DYT 18* (PED) *DYT10* and *DYT19* (PKD) *DYT8* and *DYT20* (PNKD)
Childhood-adulthood	Isolated/ combined with pyramidal	Static	Hemidystonia	**Nervous system pathology without degeneration** Lesion/ infarct/ infections
Adolescence to	Isolated/	Variable and	Variable not fitting	**Acquired**

adulthood	combined	fluctuant	into a pattern	Psychogenic

Abbreviations

AD- Autosomal dominant, AR autosomal recessive, XL- X chromosome linked. ROPD- rapid onset dystonia-parkinsonism, YOPD Young onset Parkinson's disease, PKD Paroxysmal kinesigenic dyskinesia, PNKD **Paroxysmal non-kinesigenic dyskinesia, PED Paroxysmal exercise induced dyskinesia**