Hemidystonia: a report of 22 patients and a review of the literature

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SUMMARY Hemidystonia defined as involuntary, sustained posturing of the unilateral arm, leg, and face was studied in 12 male and 10 female patients. Hemidystonia was caused by cerebrovascular disease in eight patients, perinatal trauma or childhood injury in four, head trauma and its sequelae in three, neuronal storage disorders in two, neurodegenerative disease in two, lesions after thalamotomy in two, and presumed encephalitis in one. Sixteen patients (73%) had CT evidence of contralateral basal ganglia damage, history of hemiparesis, or both. Brain damage before 7 years of age produced contralateral hemidystonia with a mean delay of 9-7 years. In older patients hemidystonia appeared within 6 months after injury. Hemidystonia may result from a disconnection between the striatum and the thalamus with relative preservation of the corticospinal pathways.

The term dystonia was coined by Oppenheim in 1911 to describe sustained posturing as well as tonic and clonic spasms of different parts of the body with muscle tone fluctuating between hypotonia and hypertonia.¹ Most patients with dystonia have primary torsion dystonia, which is either sporadic or hereditary.²⁻⁶ The biochemical and pathophysiological mechanisms of primary (idiopathic) dystonia are unknown.⁷⁻¹⁰ Dystonia rarely occurs as a psychiatric condition, although it is frequently misdiagnosed as such.¹¹⁻¹³

Dystonia has been classified according to distribution as either focal, when only a single body part is involved (torticollis, blepharospasm, oromandibular dystonia, writer's cramp, or foot dystonia), multifocal or segmental, when more than one body part is involved, or generalised, indicating involvement of at least one leg and a cranial or a brachial structure.^{3,5,6,14} Occasionally the unilateral arm, leg, and face are affected. This presentation has

aetiologic significance and calls for a separate category of hemidystonia.

We describe 22 patients with acquired hemidystonia and propose a pathogenic mechanism for this disorder.

Methods

Three hundred and nine patients with dystonia, defined as involuntary sustained posturing, have been evaluated at the Baylor Movement Disorder Clinic from 1978 to 1984. Twenty-two patients (7.1%) had dystonia of the arm and leg on one side of the body and some had involvement of the neck or face (tables 1 and 2). None had generalised dystonia, positive family history, or Ashkenazi Jewish background. Patients with focal dystonia, Wilson's disease, Huntington's disease, Hallervorden-Spatz disease, and other hereditary neurological syndromes were excluded. There was no history of exposure to manganese, carbon monoxide, drugs, or other agents known to produce persistent dystonia.^{15,16} All patients were filmed.

Results

There were 22 patients, 12 men and 10 women (table 1). The mean age at onset of hemidystonia was 36 years (range: 2 to 72 years). Eleven patients (50%) had focal neurologic deficits prior to the onset of dystonia. The mean latency between the onset of focal signs and the appearance of abnormal posturing was 4 years. Patients 1 to 6 acquired cere-

Presented in part during the 109th annual meeting of the American Neurological Association, 8-10 October, 1984, Baltimore, Maryland.

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Received 23 August 1984 and in revised form 14 November 1984.
Accepted 17 November 1984

Table 1 Historical data on 22 patients with hemidystonia

Patient No.	Age at onset of dystonia (yr) Handedness, Sex	Predisposing factors and observations	Latency between predisposing brain insult and onset of dystonia	Duration of disease at last follow-up (yr)	Associated disorders
1	29, R, F	Premature delivery by C-section, left-sided limp at 1 year, diagnosed as having cerebral palsy at 7 years, increased involuntary movements on left at 29 years	29 yr	2	Oligomenorrhoea/amenorrhoea, Mitral valve prolapse
2	2, L, M	Delivery by C-section, flexion of right arm with ipsilateral toe-walking at 2 years	2 yr	15	—
3	33, L, F	Multiple complications during maternal pregnancy, cyanotic at birth, gradual onset of cramping spasm of left hand while typing	32 yr	1	—
4	8, R, M	Gradual onset of flexion of left fingers	Unknown	7	—
5	2, R, F	Acute febrile illness at age 2 years resulting in right hemiparesis with "spasms," patient became left-hand dominant after this event	1 month	12	—
6	8, R, M	Multiple trauma at age 7 years with cardiac arrest during reparative surgery resulting in obtundation and left hemiparesis followed by flexion of left wrist and elbow	14 months	8	—
7	21, R, M	Closed head injury with loss of consciousness at age 17 years resulting in transient left hemiparesis, developed spasms of left great toe at age 21 years	4 yr	9	—
8	62, R, F	Mild head trauma with no loss of consciousness, followed by "spasms" of left hand, flexion of left wrist, and inversion of homolateral foot	4 days	2	Left hemiparkinsonism, myoclonic jerks and tremor of left arm
9	14 R, F	Normal birth and development, parents are first cousins of Lebanese Moslem origin, gradual onset of involuntary movements of right extremities	—	1½	—
10	25 L, F	Normal birth and development with no consanguinity in family, gradual onset of involuntary flexion of the left hand	—	3½	—
11	64, R, M	Gradual onset of abnormal gait with intortion of right foot	—	2½	Chronic sensorimotor neuropathy with fasciculations
12	42, R, F	Gradual onset of involuntary flexion of fingers and wrist of left hand	—	3	Parkinsonism
13	42, L, M	Right stereotaxic thalamotomy with onset of incoordination and involuntary spasms of left hand immediately after surgery	—	5	Closed head injury 1 year before onset of Parkinsonism diagnosed 2 years before surgery
14	58, R, M	Right stereotaxic thalamotomy with onset of hyperextension of left fingers and hyperflexion of left toes immediately after surgery	—	Involuntary posturing resolved in 3 months	Parkinsonism diagnosed 8 years before surgery
15	36, R, F	Sudden onset of left hemiparesis followed by involuntary "twisting" of weakened extremities	1 month	19	Left hemiparkinsonism diagnosed 17 years after event
16	61, R, M	Sudden onset of left hemiplegia with gradual resolution followed by spasms in left hand and toe-walking in left foot	6 months	10	—
17	46, R, F	Sudden onset of left hemiparesis followed by hyperextension of left fingers and flexion in ipsilateral wrist	1 month	4	Aortic stenosis, placement of Starr-Edwards valve 9 years before event
18	58, L, F	Gradual onset of flexion of left hand and arm with loss of control of ipsilateral foot in 1980	—	3	Severe headache for 3 days in 1978
19	40, R, M	Sudden onset of left hemiparesis followed by flexion of ipsilateral wrist and arm	1 month	16	Generalised tonic-clonic seizures after hemiparesis, vertebrobasilar insufficiency
20	72, R, M	Sudden onset of right hemiparesis in 1981 followed by coarse tremor of head and right arm in 1982 after bypass procedures	1 month	2	Hypertension, peripheral vascular disease, 2 aorto-femoral bypass procedures in 1982
21	22, R, M	Sudden onset of left hemiparesis and hemisensory deficit associated with headache gradual development of flexion of left wrist, flexion of metacarpophalangeal joints and extension of interphalangeal joints of fingers, flexion-extension dystonic tremor of left hand, and extension of left toes	1 month	1	Febrile seizures at age 4 years, migraine headaches
22	52, R, M	Sudden onset of headache and dysarthria from left thalamic-basal ganglia haemorrhage in 1980, followed by "twisting" movements of right foot and arm	1 month	4	Myocardial infarction in 1979, triple coronary artery bypass, bilateral carotid endarterectomies, and left retinal artery occlusion in 1983, hypertension

Table 2 Clinical characteristics of 22 patients with hemidystonia

Patient No.	Distribution of dystonia	Neurodiagnostic procedures		Therapy and results	
		CT	Other	Medical	Surgical
1	(L)AH, Fo	Right cerebral atrophy, arachnoidal cyst in right frontoparietal area	Angiogram showing avascular mass corresponding to arachnoidal cyst	Subjective improvement with Sinemet and baclofen	—
2	(R)AH, Fo	Left cerebral atrophy, left porencephalic cyst	EEG with slowing in left posterior region	Subjective improvement with baclofen and trihexyphenidyl	—
3	(L)AH, Fo	Linear area of infarction in posterior right putamen	NC	Subjective improvement with trihexyphenidyl	—
4	(L)AH, LF	Infarction in right basal ganglia	Normal cerebral angiography	No response to Sinemet, haloperidol, and ethopropazine	Stereotaxic destruction of right Vim thalamic nucleus with resumption of dystonia within 2 months
5	(R)AH, Fo	Infarction in left striatum	NC	Subjective improvement with trihexyphenidyl	Triple arthrodesis and tendon transfer in right foot
6	(L)AH, LF	Normal	NC	No response to multiple drugs	Stereotaxic destruction of ventralis oralis interna of right thalamus with transient improvement
7	(L)AH, LF	Encephalomalacia in right globus pallidus	NC	No response to multiple drugs	Stereotaxic destruction of right Vim thalamic nucleus with mild improvement of left hand dystonia
8	(L)AH, Fo	Enhancement in right posterior thalamus, empty sella	NC	No response to Sinemet	—
9	(R)F, AH, LF	Minimal cerebral atrophy	EM of lymphocytes positive for ceroid inclusion bodies	Subjective improvement with trihexyphenidyl	—
10	(L)AH, Fo	Normal	Conjunctival biopsy showing fibroblasts distended by ceroid-engorged vacuoles	No response to multiple drugs	—
11	(R)AH, LF	Normal	EMG/NCS showing sensory neuropathy, muscle biopsy showing fiber type grouping	Subjective improvement with Sinemet	—
12	(L)H, Fo	Normal	NC	Subjective improvement with Sinemet	—
13	(L)H, Fo	Normal	NC	No response to Sinemet, trihexyphenidyl, and amantadine	—
14	(L)H, Fo	Normal	NC	Resolution of dystonia following 3 months of treatment with Sinemet and trihexyphenidyl	—
15	(L)AH, LF	Infarction of right basal ganglia	NC	No response to trihexyphenidyl and benzodiazepine	—
16	(L)H, Fo	Infarction of genu of right internal capsule	NC	No response to trihexyphenidyl	—
17	(L)H, Fo	Enhancement in area of previous infarction in right basal ganglia	Angiogram showing embolic occlusion of trifurcation of right middle cerebral artery	No response to Sinemet	—
18	(L)AH, LF	Encephalomalacia in right basal ganglia	NC	No response to multiple medications	—
19	(L)AH, Fo	Infarction of right basal ganglia	NC	Subjective improvement with trihexyphenidyl	—
20	(R)AH, Fo	Normal	Palatal myoclonus documented EMG	No response to multiple medications	—
21	(L)H, Fo	Encephalomalacia in area of posterior limb of right internal capsule	Normal cerebral angiography	No response to propranolol or trihexyphenidyl	—
22	(L), AH, LF	Left thalamic-basal ganglia haemorrhage in 1980. Lucency in external capsule extending into striatum in 1983	Cerebral angiography in 1983 showing 60% stenosis of both common carotid arteries	No response to haloperidol, marked spontaneous resolution over four years	—

R = right, L = left, M = male, AH = arm-hand, LF = leg-foot, Fo = foot, H = hand, F = face, NC = noncontributory, EM = electron microscopy, EMG/NCS = electromyography, nerve conduction studies
Trihexyphenidyl = benzhexol

bral insults before 7 years of age and had a mean latency of 9.7 years (range: 1 month to 32 years) from the acute injury to the onset of hemidystonia. Adult patients developed hemidystonia within 6 months after the predisposing injury.

The mean duration of dystonia was 6 years (range: 2 months to 19 years). The patients were

followed an average of 2 years. Two patients (nos. 13 and 14) had been followed for Parkinson's disease for 2 and 8 years, respectively, before they developed hemidystonia after thalamotomy.

CT scans were performed on all 22 patients (fig 1A–B and 2B). Eleven patients (50%) had evidence of basal ganglia damage on CT (tables 2 and 3).

Fig 1 (A) Right cerebral atrophy and arachnoid cyst in patient 1. (B) Contrast enhancement in area of embolic infarction in right basal ganglia in patient 17. (C) Infarction of genu of right internal capsule in patient 16. (D) Post-traumatic encephalomalacia in the area of the right globus pallidus in patient 7.

Diffuse damage in the basal ganglia region contralateral to hemidystonia was present in seven patients (Nos. 1, 2, 4, 15, 17–19), and in 3 (Nos. 3, 5, 22) the lesions were confined to the striatal nuclei, involving chiefly the putamen. Cerebral angiography showed an avascular mass corresponding to the location of the arachnoid cyst in patient 1, an embolic occlusion of the right middle cerebral artery in patient 17, and bilateral carotid stenosis in patient 22. Electroencephalograms in nine patients were

Fig 2 (A) Patient 4. Note abduction of the left arm at the shoulder with flexion or extension of the elbow, inversion of the ipsilateral leg and foot, and dystonic flexion of the left fingers. (B) Infarction of right basal ganglia and cerebellar hypoplasia in patient 4.

Table 3 CT scan in hemidystonia (n = 22)

Location of Abnormality	N	Aetiology	N
Basal Ganglia	11		
Diffuse	7	Infarction	5
Striatum	2	Porencephalic/ Arachnoidal Cyst	2
Striatum/Thalamus	1	Infarction	2
Globus Pallidus	1	Hemorrhage	1
Internal Capsule	2	Trauma	1
Thalamus	1	Infarction	2
Generalised	1	Trauma	1
Normal	7	Atrophy	1

normal except for patient 2 who had slowing in the left posterior region caused by the porencephalic cyst. Myelograms in two patients and CSF in six were unremarkable. Light and electron microscopy of peripheral lymphocytes in patient 9 and a conjunctival biopsy specimen in patient 10 suggested a neuronal storage disorder.

Patients 2, 3, 5, 9, and 19 had subjective improvement with trihexyphenidyl in dosages up to 30 mg per day used alone or in combination with baclofen. Patients 1, 11, and 12 improved with Sinemet (carbidopa and levodopa) up to 100/1000 mg per day. In patient 1 this effect lasted only 1 year. Neurological examination revealed little or no improvement with medical treatment. Patient 14 had spontaneous remission of hemidystonia caused by thalamotomy. The hemidystonia in patient 22 gradually resolved over a 4-year period. The remaining 11 patients received no benefit from treatment with one or more of the following drugs: Sinemet (carbidopa/levodopa), trihexyphenidyl, haloperidol, tetrabenazine, reserpine, propranolol, clonazepam, carbamazepine, dantrolene sodium, orphenadrine, and clorazepate. Three patients (Nos. 4, 6, and 7) received transient benefit from stereotaxic thalamotomy.

The aetiologies of hemidystonia in the 22 patients and in 52 others reported in the literature are listed in table 4. Sixteen of our patients (72%) developed hemidystonia in association with cerebrovascular disease (8), childhood injury or presumed encephalitis (5), or head trauma and its sequelae (3). Of the remaining six patients, two had a neuronal storage disease, two developed hemidystonia in association with degenerative neurological disorders, and two became hemidystonic after thalamotomy for Parkinsonian tremor.

Discussion

All 22 patients acquired hemidystonia as a result of a structural brain lesion, a storage disease, or a degenerative neurological disorder. A pre-existing hemiparesis and evidence of striatal damage on CT scan were important risk factors for the subsequent development of dystonia.

The latency between brain injury and the onset of hemidystonia was from 14 months to 29 years in patients 1, 2, 3, and 6 who sustained cerebral insults during infancy or childhood. Mitchell suggested that the delay in onset of hemichorea or athetosis following hemiplegia was caused by progressive changes in the original brain lesion.¹⁷ Burke and co-workers have speculated that "delayed-onset dystonia" is related to aberrant neuronal sprouting in the central nervous system following a static lesion.¹⁸ Similar mechanisms are postulated in patients with blepharospasm and other facial dystonias after rostral brainstem lesions.¹⁹

Dooling and Adams examined the brains of five patients with "posthemiplegic athetosis" and found generalised gliosis of the thalamus in 1 brain and striatal damage in 4.²⁰ They suggested that any lesion capable of isolating the striatum from the ventralis anterior, centrum medianum, and ventralis lateralis nuclei of the thalamus, while preserving the corticospinal pathways, could result in contralateral involuntary movements. Although they used the term athetosis to describe these movements, a review of the case histories suggests that their patients had hemidystonia. Other reports provide additional evidence for the theory of striato-pallido-thalamic disconnection (table 4). All but five of the 35 reported cases of hemidystonia had focal lesions of the striatal nuclei by radiographic studies

Table 4 Aetiologies of hemidystonia: present series and literature review

Aetiology	Present Study (22 Patients, 1979-84) Number of patients		Literature Review (28 Reports, 52 Patients, 1937-84) References (by first author)	
Perinatal Trauma or Childhood Onset	4 [1-4]	12	[Burke (3), Denny-Brown (1), Dooling (2), Marsden (4), Oppenheimer (1), Quagliari (1)] ^{18 52 20 67 52 23}	
Presumed Encephalitis	1 [5]	4	[Dooling (3), Gordin (1)] ^{20 66}	
Head Trauma and Sequelae	3 [6-8]	14	[Andrew (1), Brett (1), Burke (1), Burton (1), Demierre (1), Maki (1), Marsden (2), Mauro (2), Messimy (1), O'Callaghan (1), Perlmutter (1), Rasmussen (1)] ^{53 54 18 55-57 67 58 59 22 24 60}	
Storage Disorder	2 [9, 10]	—	—	
Neuro-degenerative Disorder	2 [11, 12]	—	—	
Post-thalamotomy	2 [13, 14]	—	—	
Cerebrovascular disease (Infarct, Bleed, or Arteritis)	8 [15-22]	17	[Austregesilo (1), Burke (1), Demierre (2), Glatt (2), Grimes (1), Marsden (5), Obeso (1), Russo (1), Soloman (1), Sunohara (1), Traub (1)] ^{61 18 56 21 62 67 69 63-66}	
Tumor	—	3	[Narbona (1), Sciarra (1), Urechia (1)] ⁷⁰⁻⁷²	
Arteriovenous Malformation	—	2	[Marsden (2)] ⁶⁷	
	22	52		

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or at necropsy.^{18 21-23} Perlmutter and Raichle found decreased oxygen metabolism and increased blood flow in the basal ganglia of a patient with hemidystonia who had a normal CT scan.²⁴

Hemichorea and hemiballism are often confused with hemidystonia and have been attributed to contralateral, striatal and subthalamic pathology caused by infarction, haemorrhage, or metastatic tumour.²⁵⁻²⁷ In a review of 32 patients with hemiathetosis, 21 necropsy specimens were found to have destruction of the contralateral striatal or lenticular nuclei.²⁸

In our series, the most frequent cause of hemidystonia was haemorrhage or infarction in the basal ganglia (eight of 22 patients): patient 17 had embolic occlusion of the right middle cerebral artery, patient 21 became hemiparetic during a migraine attack, and patient 22 suffered a thalamic-basal ganglia haemorrhage, patient 18 probably developed hemidystonia from migrainous vascular occlusion, and the remaining four patients probably had thrombotic occlusions of arteries supplying the basal ganglia.

Dystonia is a rare complication of cerebrovascular disease in the basal ganglia territory. None of the 20 adult patients with CT-documented basal ganglia lesions reported by Naeser *et al*²⁹ and Damasio *et al*³⁰ had hemidystonia. Graff-Radford and colleagues described five patients with thalamic infarction, two of whom had hemiparesis with no associated dystonia.³¹ Posthemiplegic dystonia seems to occur more often in children than adults, possibly due to aberrant neuronal sprouting during brain maturation.^{20 32}

Sparing of the corticospinal tract and disruption of the pathways between the striatum, pallidum, and thalamus are probably essential for secondary dystonia to occur.²⁰ In our series, patients 16 and 21 had partial involvement of the corticospinal tract in the internal capsule on CT scan. All others had no apparent corticospinal involvement.

Patients 9 and 10 developed hemidystonia as a result of suspected ceroid lipofuscinosis, a heretofore unreported complication of this disease. These patients highlight the importance of a thorough search for storage disorders in all patients with atypical dystonia. Light and electron microscopy of peripheral lymphocytes or biopsy material taken from conjunctiva, skin, or rectum will usually establish the diagnosis. Sea-Blue histocytes may be seen on bone marrow biopsy.^{33 34} It is possible that these two patients will eventually develop generalised dystonia, a more typical manifestation of the storage disorders.³⁵⁻³⁸

Patient 11 presented with hemidystonia, peripheral neuropathy, and fasciculations suggesting

motor neuron disease. His family history was unremarkable. Similar examples of "pallido-pyramidal" disorders manifesting as generalised dystonia, usually with autosomal dominant inheritance, were reviewed by Gilman and Romanul.³⁹

Patient 12 had hemidystonia as a manifestation of Parkinson's disease. Dystonic postures of the hands and feet are frequently seen in patients with Parkinson's disease and with dopaminergic therapy.⁴⁰⁻⁴² Gortvai successfully treated 150 Parkinsonian patients with dystonia by stereotaxic thalamotomy.⁴³ Patients 13 and 14 developed hemidystonia following thalamotomy for Parkinsonian tremor. Patient 14 represents one of two "cures" of hemidystonia reported in this series. He has been followed for 2 years after thalamotomy with no recurrence of dystonia.

Some of our patients had no predisposing brain injury and may eventually develop generalised idiopathic torsion dystonia.^{6 14 44 45} Marsden and Harrison estimated that maximal progression of generalised dystonia may take as long as 10 years.⁴⁶ Generalised dystonia frequently begins as action-induced focal dystonia, such as occupational cramp,⁴⁷ whereas hemidystonia often starts at rest.

The response to medications was disappointing. Various muscle relaxants, tetrabenazine, and high dosage anticholinergic agents,^{6 48 49} provided minimal or no improvement in motor performance. Fahn treated three hemidystonic adults and two children with up to 50 mg of trihexyphenidyl or 800 mg of ethopropazine per day with no success.⁴⁸

Three patients underwent stereotaxic thalamotomy for treatment of hemidystonia, but obtained only mild or transient improvement. This is in contrast to reportedly successful results obtained in 12 patients with hemidystonia and 161 patients with dystonia musculorum deformans.^{50 51}

We conclude that hemidystonia is often preceded by hemiparesis and that it usually implies a structural, degenerative or metabolic lesion of the contralateral basal ganglia. It is produced by a disconnection between the striatum and the thalamus with preservation of the corticospinal tract. A delay in onset of dystonia is commonly seen in patients with acute brain damage in childhood. Treatment is often disappointing, but favourable results may be obtained with medical or surgical therapy.

We thank Dr R Nick Bryan for his review of neuroradiological studies.

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