

RESEARCH ARTICLE

100 CHILDREN WITH ACUTE ATAXIA; A SURVEY IN MOFID CHILDREN'S HOSPITAL

**P. Karimzadeh MD¹,
M. Togha MD²,
M. Ghofrani MD³**

1. Pediatric Neurologist, Associate professor, Shaheed Beheshti University of Medical Sciences, Pediatric Neurology Department, Mofid Childrens' Hospital.

2. Neurologist, Assistant professor, Tehran University of Medical Sciences.

3. Pediatric Neurologist, Professor, Head of Pediatric Neurology Department, Mofid Childrens' Hospital, Shaheed Beheshti University of Medical Sciences.

Corresponding Author:

Parvaneh Karimzadeh MD

Tel: +98 21 22227033

Fax: +98 21 22220254

E-mail: Pkarimzadeh@yahoo.com

Abstract:

Objective:

The term "Ataxia" refers to disturbances of body posture and movement that are normally controlled by the cerebellum, frontal lobes and the posterior columns of the spinal cord. The primary symptom and the most prominent feature of ataxia is abnormal gait which is characterized by lurching and wide base walking.

Ataxia was considered acute, if it had occurred within the two preceding weeks. Knowing how frightening acute-onset Ataxia is for the family is not surprising that the condition prompts an immediate visit to the physician.

Material & Methods:

In view of the lack of information in our country, on the etiology of sudden-onset Ataxia, the authors enrolled 100 children with the chief complaint of acute loss of equilibrium, who came to the attention of the Pediatric Neurology Department over a two year duration (Sept.2001-Sept 2003); they were admitted to the Mofid Childrens' Hospital and all necessary investigations were carried out.

Results & Conclusion:

The results revealed that Acute Cerebellar Ataxia was the most common cause of the problem, the second most frequent being drug intoxication, which most commonly occurred in patients, 2-4years old. The remaining causative factors in order of descending frequency consisted of infectious polyneuropathy, migraine, opsoclonus-myoclonus, brain tumor, acute disseminated encephalomyelitis, multiple sclerosis, and epilepsy.

Key words: Acute ataxia, Children, Acute cerebellar ataxia

Introduction

The term "Ataxia" is used to refer to disturbances of the body's posture and movement normally controlled by the cerebellum, frontal lobes and the posterior columns of the spinal cord. The primary symptom and the most prominent feature of ataxia is abnormal gait characterized by lurching and wide base walking. The ataxic gait is wide-based, associated with lurching and staggering and disturbing to the onlooker, creating a fear that the patient may fall.

When the pathology is located in the vermis of the cerebellum, the child's body constantly moves to and fro and head bobs (titubation), as the individual tries to sit still; however if the lesion has its origin in the cerebellar hemisphere the patient has a tendency to fall ipsilaterally. Whenever the pathology affects the peripheral nerves or posterior column, disturbances of sensory input to the cerebellum, make the person look constantly at their feet to ensure his/her stability and to prevent falling. The situation becomes most difficult when the individual closes his or her eyes, which causes the patient to fall to the floor.

Etiology of acute ataxia differs vastly from that of a child who suffers chronic or progressive ataxia. Drug intoxication, and post infectious cerebellitis are the two most common causes of sudden onset ataxia in the pediatric age group, among children who previously enjoyed healthy life (1,2,3,4,5,6,7,8). The causes thereafter encountered were migraine, encephalitis, genetic disorders (Episodic ataxia type 1 and 2, Hartnup disease, Maple syrup urine disease) and some post infectious immune diseases (Miller fisher syndrome, Multiple Sclerosis, Myoclonic encephalopathy and Neuroblastoma), ataxia due to Epilepsy, vascular disorder (Kawasaki) and brain tumor (1,2,3,5,8,9,10,11,12,13,14,15).

Being aware of acute ataxia as an illness that immediately brings the affected child to the pediatrician or child neurologist's attention, and considering the lack of any data obtained from related surveys in this field in our country, we decided to investigate the etiology of acute ataxia in children brought to the pediatric neurology department of the Mofid Children's Hospital in Tehran.

Materials and methods

Our patients consisted of 100 children, referring to the pediatric neurology department of the Mofid Children's hospital, over a 2 year period (Sept 2001-Sept 2003), with a chief complaint of acute ataxia, which had appeared within the last two weeks; all the children were admitted and the appropriate investigations were carried out. Children with chronic static or progressive ataxia were excluded from the study. The findings were analyzed by SPSS and statistical software.

Results

Of a 163 patients brought to the department, the 100 (42 girls, 58 boys; table I) who had developed the condition, i.e. chief complaint of ataxia, were enrolled; the following results were obtained:

Table I: Age distribution of patients with acute ataxia

age	Sex		Total
	female	male	
under 1 year	–	–	–
1-2 years	10	15	25
2-5 years	10	14	24
5-10 years	18	21	39
above 10 years	4	8	12
Total	42	58	100

The youngest and oldest patients were 15 months old and 13 years respectively (table I). 51 patients showed history of previous febrile illness, of which in 14, fever appeared one week prior to ataxia; in 28, the onset of

fever was 1-2 weeks before and in 9 cases the patients problem arose to 2-3 weeks following onset of fever (table II).

Table II: Duration between previous febrile illness and acute Ataxia

Sex Time	Female	Male	Total
Before 1 week	4	10	14
1-2 week	11	17	28
2- 3 week	3	6	9
Total	18	33	51

42 cases had other features of cerebellar dysfunction like finger to nose and heel to shine, dysmetria , etc. 26 cases simultaneously had dysarthria or scanning speech with ataxia. 17 cases had hypotonia; in the upper limb we considered hypotonia with scarf sign (in infants) and pronator sign (in children); in the lower extremity the

fall away test (in infants) and barre sign (in children) were taken into account. 51 cases had histories of previous febrile illness, including chicken pox (11 patients), mumps (one patient), herpes simplex viruses (one patient), and nonspecific febrile illness (38 patients), (table III).

Table III: Type of infection in A.C.A

Type	Number
Chicken Pox	11
Mumps	1
Herpes Simplex V.	1
Non specific febrile illness	38
Total (A.C.A)	51

In 22 patients, ataxia was attributed to drug toxicity; in 17 of these, drugs found to be the cause of ataxia were Diazepam -3, Antihistamine-4, Phenytoine-2, Lorazepam-3, Oxazepam-, and Phenobarbital-3; in 5 however, the nature of the drugs could not be identified. Among these 22 patients, 20 children were between 2-5 years old, who had ingested the drugs accidentally; the remaining 2 patients were epileptic and their ataxia was due to an overdosage of anti epileptic drugs.

Drug toxicity in one 10 year old patient, was due to Phenobarbital and in another 11 year old toxicity was secondary to phenytoin. This was confirmed by drug level measurement. 5 patients in these series were finally diagnosed as migraine cases; their characteristics were: One had a positive family history of migraine and a

history of motion sickness having sudden onsets of acute ataxia whenever he rode in a rotary machine, e.g. merry-go-round, in amusement parks. Patient two, had repeated episodes of severe confusion, occurring 48 hours prior to ataxia onset; all the investigations including neuroimaging and EEG were normal, and this patient improved remarkably when propranolol was administered. Patient 3, a 5 year-old, had acute ataxia after minor head trauma; his condition normalized completely after forty-eight hours of rest. In patients 4 and 5 the chief complaint was of headache, vomiting, swallowing difficulty and ataxia. Various investigations were normal, and their symptoms disappeared in a short period of time, we presumed they were suffering from basilar migraine (Table IV).

Table IV: The etiologies of acute Ataxia

Etiology	Number
A.C.A*	51
Drug intoxication	22
Post infectious poly neuropathy	12
Migraine	5
Opsoclonus myoclonus,	5
Brain Tumor	2
Acute disseminated encephalomyelitis	1
Multiple Sclerosis	1
Epilepsy	1
Total	100

(*Acute Cerebellar Ataxia)

In 2 patients the cause of acute ataxia was brain tumor; these two children had papilledema in fundoscopic examination and neuroimaging study revealed space occupying lesions in the posterior fossa and frontal lobe (table IV). One ataxic child, whose ataxia was preceded by febrile illness, had elevated levels of protein in his C.S.F examination, increased deep tendon reflexes, and right abducens nerve palsy. M.R.I showed increased signal on T2 weighted, particularly at the gray and white matter junction, characteristic of A.D.E.M. (15); this child had suffered from a febrile illness for one week duration, and three days after the fever subsided the child became ataxic. This patient improved with intravenous corticosteroid therapy (table IV). One 15 year-old girl presented with acute ataxia, lower and upper limb weakness and visual impairment; her fundoscopic examination showed papillitis and the ophthalmologist found central and cecentral scotoma and visual field defect; cranial M.R.I. showed M.S. plaque.

She showed clinical improvement following administration of intravenous corticosteroid (16) (table IV). One child, a known case of symptomatic epilepsy who had been confused for the last 3 days, came to us with the complaint of ataxia; he had had repeated episodes of generalized tonic clonic convulsion in the past. According to the parents, the child was having repeated staring spells during the previous 10 days. At the time of admission, he was quite disoriented regarding time or place, and his E.E.G showed continuous spike wave discharge. Diagnosed as "absence status", he was placed in I.C.U and diazepam drip therapy was instituted, leading to complete recovery from ataxia and staring spells within 72 hours (table IV). 5 cases of ataxia, age range 14-27 months, presented themselves with opsoclonus-myoclonus; all 5 cases had ataxia, dancing eye movement and myoclonus in both upper and lower limbs as their presenting symptoms. In 4, investigation results for neuroblastoma, including chest and abdomen M.R.I and sonography, and V.M.A.-H.V.A assessments of 24 hours urine were normal (10,11,12,13). These 4 patients had full recovery in response to intravenous corticosteroid therapy. In the fifth case, investigations confirmed a diagnosis of occult neuroblastoma and case was referred for management to the oncology department (table IV).

In 12 of the children, we made a conclusive diagnosis of Guillaine Barre and Miller Fisher syndromes (G.B.S in 10 and Miller fisher syndrome in 2). Although the presenting symptom in the 12 cases mentioned was ataxia, their lack of equilibrium was in fact secondary to muscle weakness. We detected the known triad of acute ataxia, areflexia and ophthalmoplegia in two of our Miller Fisher cases. C.S.F. examination confirmed albuminocellular dissociation and electrodiagnostic study was suggestive of acute peripheral neuropathy. All of these 12 patients showed clinical improvement following intravenous immunoglobulin therapy (table IV).

In literature there are reports of acute ataxia secondary to the conversion reaction, channelopathy, metabolic diseases (intermittent M.S.U.D, Episodic ataxia 1,2, Hartnup ,...), posterior fossa hemorrhage ,etc.; however none of 100 patients studied in our investigation had any of the above mentioned conditions as etiology of their acute ataxia(1,2,3,4,5,6,7,8,9,14) (table IV).

Discussion

Our study revealed that in 51 patients, acute cerebellar ataxia was the cause of sudden onset ataxia; in all of them, the condition was preceded by febrile illness, this being the most common case of the condition. Of the 51 cases with this diagnosis, 11 cases suffered from chicken pox. Herpes Simplex and mumps viruses encephalitis each caused ataxia in one patient respectively.

In the remaining 38 patients, sudden onset ataxia was preceded by a febrile illness for which we were unable to pinpoint the infection causing agent. The majority, 37 cases, had had their febrile illness 1-3 weeks before the occurrence of acute ataxia; 14 patients had a previous infection less than one week prior to their illness.

In our investigation, we observed Varicella to be the most common visible entity that we could clinically determine as the cause of ataxia, a conclusion in agreement with other studies(1,2,3,4,5,6,7,8,9). The second most frequent cause of the problem under discussion was drug intoxication occurring in 20 children, aged 2-4 years; this finding is also confirmed by other reports (1, 2, 3, 4, 5, and 6). In the present study, infectious polyneuropathy was found to be the third prevailing cause of the acute ataxia found in 12 patients (8).

The remaining etiologic factors in order of descending frequency consisted of migraine, Opsoclonus-myoclonus, brain tumor, multiple sclerosis, acute disseminated encephalomyelitis, and epilepsy (1,2,3,4,5,6,7,8,9,10,11,12,13,14,16).

At the time of the writing this paper, the patients had been followed for at least 2 years. Children who had previously suffered from acute cerebellar ataxia, drug intoxication, infectious poly neuropathy, migraine, MS, A.D.E.M, and 4 patients of opsoclonus myoclonus, remained asymptomatic and had no recurrence of the problem (3,10,11,13). Findings of other studies indicate that full gait recovery took under 2 weeks on average, and the longest duration of neurological sign was 2-3 weeks; our findings were in accordance to these results (1, 2,3,4,7,8).

Of two patients with brain tumors, one expired after surgical intervention and the other is under care of a neurosurgeon. The case of neuroblastoma is being managed by a pediatric oncologist (10, 11, 12, and 13).

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