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The Early Treatment of A Boy From Virginia With Ataxic Cerebral Palsy

Aamir Jalal Al-Mosawi

Advisor doctor in pediatrics and pediatric psychiatry, Baghdad Medical City and Iraqi Ministry of Health, Baghdad, Iraq

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***Corresponding author:** Aamir Jalal Al-Mosawi, Advisor doctor in pediatrics and pediatric psychiatry, Baghdad Medical City and Iraqi Ministry of Health, Baghdad, Iraq

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Abstract

Background: Ataxic cerebral palsy is a very heterogeneous chronic disabling neurological disorder associated with significant motor impairment and disability. It can be caused by congenital abnormalities, birth asphyxia and kernicterus. There are no specific therapies, but we have previously reported with use of novel multi-factorial therapies with benefits in patients with ataxic cerebral palsy. The aim of this paper is to describe the early treatment of a boy from Virginia, USA who had ataxic cerebral palsy.

Patients and methods: The boy had significant ataxic disability and was unable to maintain a stable posture when seated on a chair and standing because of the ataxic movements. He was still unable to stand holding furniture, but he was trying to step when his mother holding him in the standing position, and taking his weight.

Results: After, one month of treatment, the mother reported improvements in all areas of disability. She described him as definitely more comfortable sitting on a chair, and never fell out or got stuck in one position. The mother reported that his supported walking was more controlled, with better balance, feet stay more flat, and he didn't bend his knees much nor nearly fell. The boy was able to stand holding furniture for about 15 seconds.

Conclusion: The early treatment of this patient with ataxic cerebral palsy was associated with obvious benefits that have not been reported with other therapies before. A more lengthy treatment course is suggested.

Key words: ataxic cerebral palsy; multi-factorial therapies

Introduction

Cerebral palsy is a chronic non-progressive encephalopathy, which is a generalized disorder of cerebral function. The cerebral disorder in cerebral palsy which results from injury to the developing brain leads predominantly to motor dysfunction associated with abnormalities of posture and movement. Cerebral palsy is not a single disease, but a heterogeneous group of disorders associated with brain damage that can be caused by a variety of congenital and postnatal cause including birth asphyxia, congenital and developmental defects, and genetic factors. Although, emphasis has commonly been made that the nature of the lesion in cerebral palsy is non-progressive, mostly to distinguish it from disorders associated with progressive brain degeneration, emphasis has although been made that in the developing child, that is untreated, the clinical picture will not seem static. The area of brain damage determines the neurological of postural and movement abnormalities which can be of the hypertonic type, type, ataxic type or mixed type. The hypertonic type includes the common spastic type and the uncommon type associated with cogwheel or lead pipe rigidity or spasticity rather than spasticity. There is no true hypotonic type, but a hypotonic stage can be the initial presentation of spastic cerebral palsy, and also ataxic cerebral palsy. The extent of the damage to the motor cortex of the brain divides the spastic forms of cerebral palsy into localization types which include:

Spastic diplegia with main involvement of the legs with associated with spastic adduction of the hips which cause scissoring, and equino-varus spasm of the feet which cause tip toeing. Brain imaging in children with spastic diplegia may show periventricular leukomalacia.

Spastic hemiplegia with main involvement of the side of the body (leg and arm) contralateral to the site of the damage, and the arm being more affected. Brain imaging

in children with spastic hemiplegia may show atrophic cerebral hemisphere with dilated ventricle contralateral to the hemiplegic limb.
Spastic quadriplegia with involvement of all limbs reflecting more severe and extensive brain damage which increases the likelihood of associated seizures and cognitive disabilities.

Double spastic hemiplegia with involvement of both sides of the body with arms more affected than the legs.

The ataxic type results from damage or defect in the cerebellar region which is clinically associated with hypotonia, weakness and other cerebellar abnormalities such as lack of coordination and tremor.

Dystonic/ athetoid type is associated with irregular involuntary movements of muscles. Athetosis is the common abnormal movement associated with this type. It is a slow purposeless muscle movement associated with extensor spasms [1-8].
The aim of this paper is to describe the early treatment of a boy from the State of Virginia who had ataxic cerebral palsy caused by kernicterus.

Patients And Methods

The patient was a boy who was delivered by normal vaginal delivery on the 29th of March 2016, after an uneventful pregnancy, and he cried at birth and didn't need resuscitation. He was healthy at birth, but was hospitalized at the age of five days because of the development of jaundice with bilirubin level above 30 mg/dL. Hyperbilirubinemia was caused by isoimmunization resulting from ABO-Rh blood group incompatibility. Mother's blood group was AB negative, and father's blood group was A+. The boy was treated with exchange transfusion and phototherapy. He also received blood transfusion to correct anemia. He developed short seizure and received phenobarbital for six months.
The mother thought that the boy was recognizing her face and showing social smile early during infancy, but he experienced delayed speech and motor development. He has been able to sit on a chair shortly after the age of one year. He started physical therapy few weeks before the age of three years. He started crawling and saying few words at the age of three years. Further, physiotherapy didn't result in obvious motor development other than making crawling easier.

Few weeks before completing the fourth year, he showed some speech development, and his speech became clearer to his mother, and started using short sentences during January, 2020. During, January, 2021, the mother was satisfied with his speech as he was speaking in full sentences, but was frustrated that the stem cell therapy which he received in Panama during the year 2020 didn't result in ant motor improvement.

The mother thought that her child didn't have significant cognitive impairments, and he had an amazing memory. She was proud that he could play by himself for an hour or two with his toys and watching movies on television (Figure-1A), and without needing anything. She also emphasized that his ability to be on his knees, and not stuck laying down has improved his quality of life (Figure-1B). The mother also thought that her son had normal emotions and displayed normal social interaction as he loved his family and friends, and he was engaging in typical arguments with his 9-year old sister and his 3-year old cousin, but nothing was out of the ordinary.

Figure-1A: The boy was playing by himself for an hour or two with his toys and watching movies on television

Figure-1B: The patient was able to sit on his knees in rather an abnormal posture

Before starting treatment (Height: 42 Inches, Weight: 37 lbs) which was started early during March, 2021:

1-He was still unable to maintain a stable posture when seated on a chair because of the ataxic movements (Figure-1C).
2-He was unable to feed self with spoon, but could feed himself small pieces of food that are not hard with his right hand. He was still unable to drink with a cup or to eat with spoon, and he had difficulties with drinking from a bottle because of the ataxic movements and lack of coordination (Figure-1D).
3-He was still unable to stand holding furniture, but he was trying to step when his mother holding him in the standing position, and taking his weight (Figure-1E).
4-He was still using napkins.

Figure-1C: Before treatment, patient was still unable to maintain a stable posture when seated on a chair because of the ataxic movements

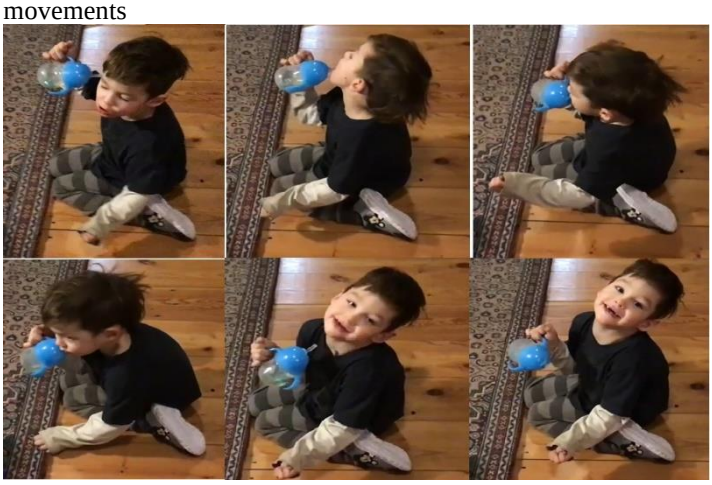

Figure-1D: Before treatment, the patient was unable to feed self with spoon, but could feed himself small pieces of food that are not hard with his right hand. He was still unable to drink with a cup or to eat with spoon, and he had difficulties with drinking from a bottle because of the ataxic movements and lack of coordination

Figure-1E: Before treatment, the patient was still unable to stand holding furniture, but he was trying to step when his mother holding him in the standing position, and taking his weight

The first one-month treatment course:

1-Intramuscular cerebrolysin 3 ml given during the morning hours, every third day (10 doses over 1 month).

2-Intramuscular citicoline 3 ml (375mg) given during the morning hours, every third day, 10 doses over 1 month.

3-Nutritional support provided as oral royal jelly 500 capsule, one capsule orally daily in the morning.

Cerebrolysin and citicoline injections were given on different days; Day 1 (Cerebrolysin), Day 2 (Citicoline), Day 3 (No injection)

Results

The patient first one-month treatment was completed on the 8th of April, 2021. Initially, the boy showed improvement with fine motor skills and was trying to draw a circle with use of chalk (Figure-2A).

Figure-2A: Initially, the boy showed improvement with fine motor skills and was trying to draw a circle with use of chalk

The mother reported improvements in all areas of disability. She described him as definitely more comfortable sitting on a chair, and never fell out or got stuck in one position (Figure-2B).

Figure-2B: The mother described the boy as definitely more comfortable sitting on a chair, and never fell out or got stuck in one position

The mother described his supported walking as more controlled, with better balance, feet stay more flat, and he didn't bend his knees much nor nearly fell (Figure-2C).

Figure-2C: The mother described his supported walking as more controlled, with better balance, feet stay more flat, and he didn't bend his knees much nor nearly fell

The boy was able to stand holding furniture for about 15 seconds (Figure-2D).The mother also reported improved speech with use of longer sentences which resulted in better communication, and the explaining things with detail, and was able to have fuller conversations.

Figure-2D: The boy was able to stand holding furniture for about 15 seconds

Discussion

Ataxic cerebral palsy is a heterogeneous chronic disabling neurological disorder associated with significant motor impairment. It has a variety of causes including congenital abnormalities, birth asphyxia and kernicterus. It can also be associated with cognitive and visual impairment and abnormal speech. There are no specific therapies, but we have previously reported with use of novel multi-factorial therapies with benefits in patients with ataxic cerebral palsy [9, 10, 11].

Imamura et al (1992) reported brain imaging findings of five patients with ataxic cerebral palsy. The first patient and her mother had an early-onset hereditary non-progressive cerebellar ataxia syndrome with spastic diplegia. The patient had atrophy of cerebellum (especially in anterior superior part) and slight atrophy of pons on brain MRI. The second patient also had MRI evidence of cerebellar atrophy especially in anterior superior part. The third patient was a boy who had CT-scan and MRI evidence of generalized spino-ponto-cerebellar atrophy and he also had dwarfism and cataracts. The boy was considered to have Marinesco-Sjörgren syndrome. The fourth patient had CT-scan and MRI evidence generalized spino-ponto-cerebellar atrophy. The fourth patient had head nodding and nystagmus and MRI evidence of MRI of cranium bifida and agenesis of anterior medullar velum.

The report of Imamura et al emphasized that ataxic cerebral palsy is a heterogeneous disorder that can be caused by very different conditions [12].

There are no specific therapies, but we have previously reported with use of novel multi-factorial therapies with benefits in patients with ataxic cerebral palsy [9, 10, 11].

Cerebrolysin is a safe mixture of active brain neuro-peptides having well-known neuroreparative properties that can contribute in improving brain functions in a variety of neurologic conditions. Cerebrolysin contains free amino acids (85%) and 15% biologically active low molecular weight amino acid sequences which include low molecular weight neuro-peptides [13]:

1-Brain-derived neurotrophic factor.
2-Glial cell line-derived neurotrophic factor.
3-Nerve growth factor.
4-Ciliary neurotrophic factor.

Cerebrolysin has been increasingly used in the treatment of a variety of childhood neurological and psychiatric disorders including brain atrophy, cerebral palsy, kernicterus, and agenesis of the corpus callosum, idiopathic mental retardation, pediatric juvenile spinal muscular atrophy, Charcot Marie Tooth disease, myelomeningocele autism, and Rett syndrome [13-18].

Citicoline is the generic name of cytidine 5-diphosphocholine (CDP-choline, cytidine diphosphate choline) when used as an exogenous sodium salt. Cytidine diphosphate choline is a mononucleotide made of ribose, pyrophosphate, cytosine and choline. It is a water-soluble naturally occurring substance that is generally grouped with the B vitamins. It is also considered a form of the essential nutrient choline. Citicoline is a safe substance with generally minor side effects which may include digestive intolerance after oral administration. Citicoline has become available throughout the world and recently it has become available in the United States as a dietary supplement [19].

Like cerebrolysin, citicoline been increasingly used with benefit in treatment of childhood neuro-psychiatric disorders including mental retardation, pervasive developmental disorders including Rett syndrome , brain atrophy , and spastic cerebral palsy [19-23].

Conclusion

The early treatment of this patient with ataxic cerebral palsy was associated with obvious benefits that have not been reported with other therapies before. A more lengthy treatment course is suggested.

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