DIAGNOSIS AND PROGNOSIS OF CEREBRO-CEREBEL-LAR DIPLEGIA

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Congenital cerebro-cerebellar diplegia is the term used for a combination of symptoms dependent upon cerebellar agenesis or to some form of injury to the cerebellum, at the time of or before birth, the exact nature of which has not yet been determined. The disorder presents varying types of symptoms which may be grouped roughly into those of flaccid and flaccid-spastic palsy, shading into the general mildly spastic state. The latter associated with cerebellar symptoms has been carefully described by Batten as congenital cerebellar ataxia. The severest type of cerebro-cerebellar diplegia is embraced in my group of cases described four years ago. In the pure cerebellar ataxia type there are hypotonia, dysmetria, gross incoordination, astasia, abasia, dysarthria, occasionally dysphagia and often complete inability to sit up because of extreme trunk ataxy. In the extreme involvement of the fore-brain the foregoing cerebellar symptoms are present together with mutism and the lowest grade of idiocy. In this latter group type there may or may not be evidence of involvement of the pyramidal tracts. In the mixed types of the association syndrome (cerebro-cerebellar type), there may be either a slight degree of spasticity in certain parts combined with hypotonia, or flaccidity in other parts of the body.

The first clinical recognition of the syndrome without recognizing the essential cerebellar character of it is to be found in descriptions of cerebral palsies, such as that of Freud, in which there were cases described as showing flaccidity, etc. As stated before, the first effort to take the cerebellar symptoms, that of the asynergies, out of the ordinary picture of cerebral palsies, was performed by Batten in his essays.
The ordinary flaccid type is fairly illustrated by the following clinical picture:

Case I is a girl of 6 years, a twin, born at the mother's first pregnancy. The birth was thought to be three weeks premature. She weighed 4½ pounds. The twin sister weighed 3½ pounds. Two subsequent children (boys) in the fraternity weighed 8½ and 10½ pounds respectively. Our little patient had an umbilical hernia, which disappeared without operation. She was an extremely passionate child, and her training has been about equally divided between conduct and social training and motor training. She was not able to sit up at the time the other twin was walking. Prior to this observation nothing unusual was noticed. The flaccid musculature of the back caused the parents to visit an orthopedic surgeon; she was then given an elaborate system of training by removable braces. She commonly lay on her back; the lower extremities bent considerably at the knees and hips strongly abducted and rotated outwardly. The legs could be easily over-extended and remained so without position. It was soon noted that the musculature of the whole body was flaccid and incapable of ordinary motivation. There was no atrophy or electrical changes. Further examination showed there was no muscular resistance and the different segments of the limbs could be maintained easily in very arbitrary positions without pain. This extreme hypotonia had naturally an intimate relationship to the absence of voluntary motion and station. At the end of two years the back muscles were sufficiently "hardened" so she was able to sit up, and was pronounced cured. At this time, when sitting and standing were encouraged, a cerebellar ataxy was noticed and incoordination of all extremities was marked when any movement was undertaken. At four years of age, some eighteen months ago, she was able to walk, but did so with the most remarkable ataxy and incoordination, comparable to a mechanical doll. All movements were dysmetric and arhythmic. Articles which she picked up would fly out of her hand as though catapulted from her fingers. She walked best with a wide base; even then she could hardly enter an ordinary doorway without striking herself on either or both sides of the door jam. In spite of the fact that she had the repellant leer of a low-grade mental defective and could not talk, by ordinary performance tests she showed no di-
minution in native ability so far as it was possible to judge. At the present time, eighteen months since the first examination, it is obvious that the ataxy, incoordination, dysmetria and hypotonia are steadily diminishing. The child is able to speak a fair number of sentences, although the speech appears to be without conscious direction. She is only learning slowly "how" she speaks. She is surprised to hear the sound of her own voice, and only after a great deal of practice is she able to produce this larger group of formerly spontaneously produced sentences. With a little assistance she feeds, dresses and undresses herself, and engages in all the ordinary everyday activities. Her general intelligence is that of her years. Her interests are far beyond her general appearance. There is even more discrepancy in her general appearance, and her performance ability than that often noted in cerebral diplegics—one is often surprised at the degree of general intelligence such children really possess.

In brief, we have here a case of cerebro-cerebellar diplegia in which the more prominent defect is cerebellar in character. Aside from the delay in ability to speak and a slight retardation in the development of the mental processes, the cerebral defect bids fair to be overcome within the next few years of training. It is in such cases one may expect the training-out of cerebellar defects in the motor asynergies to give the most brilliant results in mental development.

The milder grade of the spastic type of this disorder is shown in the following:

Case II is that of a little girl now eleven years old. She was brought to me first when seven years of age, suffering from epileptiform convulsions and a "peculiar lack of coordination." A third and a second maternal cousin were epileptic. A maternal aunt was neurotic and subject to depressions. The mother and her brother were not of a nervous type, but were very slow in games and sports which required agility.

Our patient was the first child in a family of two. Her birth was beyond term (2 weeks); there was a face presentation, and delivery was instrumental. She was a blue baby, and the cord was about the neck. She weighed 7½ pounds. She was not a particularly passionate child, but exhibited a few tantrums. She was supersensitive in regard to taking food and becoming accustomed to
clothing. Dentition was slow and difficult. She was constipated at times, but had no convulsions until five years of age. At about two years it was noticed she was backward in walking. She had only been able to stand with difficulty at 12 months, and this was maintained with much wavering. There was some rigidity in the right leg and arm, and a slight degree of flaccidity on the left side, especially in the leg.

Our patient had a double internal strabismus, which was more marked on the left. The epileptic attacks were not classic; there was no tongue-biting or voiding. They were usually in the left arm, focal in onset, although at severer periods they became general attended by loss of consciousness, and a short coma followed. There have been no attacks for the past four years.

At the time she came to me for examination at the age of seven, psychometric tests showed a mental retardation of a little less than two years. This retardation has been lessened somewhat in the past four years. Examination showed a fairly well developed child for her age, in good physical health. There was an irregular implantation of the teeth. The palate was low, with broad torus. Swallowing was difficult. The speech was indistinct, inclined to be monotonous and of the scanning type. She was markedly inco-ordinate in all movements. A marked ataxy was shown in standing and walking; marked dysmetria was present also in the upper extremities. Attempts to run increased the incoordination. The reflexes were normal. A general neurological examination was otherwise found to be negative.

In the four years intervening between the first examination and the present time, our patient steadily gained in physical and mental development. The dysmetria has greatly disappeared; the incoordination and ataxy are only markedly noticeable during excitement and fatigue; when overtired she is quite awkward, and the right leg shows considerable spasticity. A slight degree of spasticity is present in the right leg when she is at rest. Attempts at rapid coordination of different segmental parts of an extremity are performed poorly, and are very irregular (adiadokokinesis).

This case is one of cerebro-cerebellar diplegia in which the cerebral involvement has much improved inasmuch as careful teaching has permitted a wider range of manual training to be applied to
the cerebral education per se. It is a good clinical illustration of Batten’s type.

Undoubtedly many more cases of congenital cerebro-cerebellar diplegia of both mild and severe types pass unrecognized in the general designation of cerebral palsy, where gross incoordination, defects of speech, and locomotor defects loom rather large in Little’s syndrome as well as in other descriptions of cerebral disorders entailing either feeblemindedness or diplegia, or both. The automatic and natural outcome of mild grades of cerebro-cerebellar defect is fairly illustrated in the following:

CASE III is a man 32 years old, single, and is preparing to reenter a medical school in which several years ago he contracted a mental torticollis, and because of which he had discontinued medical study. He was born after an instrumental dry delivery. He was a blue baby for several hours. Physical development was delayed, and he did not talk and walk until the middle of his second year. He then walked with an uncertain straddling gait. Various devices were tried to overcome the weakness in the legs. There was a marked spasticity in the left leg, but a mild degree of hypotonus in the right. The spasticity and flaccidity, awkwardness and incoordination were slowly overcome, and at the age of sixteen it was fairly trained out by an elaborate system of physical gymnastics. His speech, which was at first delayed, was scanning, syllabic and over-precise, and this still endures in greater part. The speech defect, the physical awkwardness, slowness and arhythmic jerkiness of segmental movements of the extremities are more than vestiges of the former cerebro-cerebellar diplegia. As a child of three or four, he could not perform any of the complicated or difficult coordinated acts, and had a “tremor” in the hands—all of which became marked whenever he became excited. His rate of mental acquirement was similar to that which he showed physically, and if his mental development depended upon rapid physical coordination, it was further handicapped. Recognizing these gross physical and mental faults, the boy was encouraged by his father to take up all sorts of physical sports, which he did. In time he became an expert long distance bicycle rider.

One would at first think this man was of the simple minded class, but on closer observation one finds he is only naive and peculiar,
somewhat lacking in discrimination and judgment, and possesses a general defect in the essential understanding of life. He is not agile mentally, nor easily adaptable to different social situations. There was a retardation in mental development all through his adolescence, but the very persistence with which he now studies and strives to win his belated goal—a medical education—shows the essential primary endowment of native ability was not defective.

The prognosis of all these varying types depends, of course, upon the combination of symptoms presented. It may be fairly stated that when the forebrain is damaged to such an extent that the mental state is no longer to be classed as retardation, but shows mental arrest or marked imbecility, such children usually never recover either from their ataxia or from their defect in mental development. Inasmuch as the prognosis in an individual case of the less severe types depends upon our being quite sure that the mental state is one of simple retardation or mental arrest, much of the definiteness of prognosis cannot be determined until one has made a very decided effort to train out the cerebellar symptoms or force the forebrain to take up vicariously the functions of the cerebellum. This being the case, the kind and character of treatment is of great importance.

While the principles of treatment really embrace mental as well as motor training, the primary point of approach in any treatment must be directed toward overcoming the motor defects. This may be embraced under general headings; first of all, the removal of the segmental incoordination. In such treatment the child is taught to appreciate by actual manipulation the flexion, extension, abduction and adduction of the different segments of the extremities. The training is not so very dissimilar to that of Frenkel's system for tabetic ataxia, and his principles for training trunk coordinations are of the utmost moment in these cases. However, when one has obtained results in removing the segmental defects, there still remains the general motor training of using these simple, isolated contractions of segments into purposive acts in which all the segments of an extremity are coordinated harmoniously. When this has been fairly well inaugurated, one may undertake more definite principles of motor training, such as are laid down in the Montessori, Seguin and kindergarten systems, with music, rhythmic games, and
sports, and under the guise of play the child is further taught to
develop coordination.

In the majority of cases there is an ataxia of speech which needs
close consideration. This may range all the way from a state of
over-preciseness of syllabication to a marked dysarthria as seen in
dissemintal sclerosis or in the gibberish of the aphasic. The process
of training here must be similar to that used in teaching the deaf
and dumb. With this motor incoordination fairly taken care of
or removed, there still remains to be handled many of the tempera-
mental defects which almost invariably go with these cases. This
fault makes these children very difficult to handle. The disciplinary
system of nursery ethics has much to do toward helping the child
to develop out of his motor and mental disorder. This type of
training is similar to that which I have laid down for epileptic
children.\

In conclusion it may be said that the essential principle of train-
ing for these children is to use (1) a general training in games,
sports, and a broad concrete system of physical and mental educa-
tion; and (2) at the same time employ a special training in physical
gymnastics to teach the use of segmental movements of all sorts,
and finally (3) to give the child a thorough understanding of the
rhythmic continuity of all the different segmental movements com-
bined in a full purposive act, be it simple or complex, or fully coor-
dinated with other bodily acts (diakonesic).

The training should be carried out for a number of years under
the supervision and direction of a capable trained nurse-teacher,
upon whom the success of the work must largely depend. I trust
these general remarks upon the training system which I have em-
ployed on a series of cases for the past four years may not only be
of service in this type of cerebellar disorder of children, but that
it may serve some purpose in encouraging a more persistent and
careful training of many children suffering from cerebral palsy
and the simpler types of feeblemindedness, where cerebellar symp-
toms of incoordination which are comparable to those most ex-
quisitely shown in these cerebellar diplegics may also be found to
exist. Perhaps the principles here laid down may be found of
service even in adult cerebellar disorders in which similar symp-
toms are in evidence if the lesions in such diseases are not too destructive or too rapidly progressive in type.

REFERENCES


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