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INFANTILE CEREBRO-CEREBELLAR DIPLEGIA, OF FLACCID, ATONIC-ASTASIC TYPE *

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INTRODUCTION

It is my purpose to place on record, for the first time so far as I am aware, a comprehensive description of a type of cerebral palsy of childhood, which I presume to name "Infantile Cerebro-Cerebellar Diplegia." Various writers have commented on the condition under the designation of cerebellar or flaccid diplegia, and very recently Förster¹ has written a paper on the condition under the head of flaccid diplegia and has given some very excellent case descriptions. Inasmuch as there has been no attempt as yet to couple this flaccid syndrome with a definite pathogeny or morbid pathologic anatomy, this paper in this latter respect may be of interest to those engaged in the study of cerebral palsy of children.

It would seem that there are two reasons for the inadequate attention which flaccid diplegias of infancy have received at the hands of clinicians. First, the spastic parietic syndrome (Little's type) has been thought to be synonymous with cerebral diplegia and cases showing any state of musculature other than spasticity, or not in accordance with this syndrome were excluded from the clinical picture of infantile cerebral palsy; second, all cases of cerebral palsy dating from birth or childhood in which flaccidity obtained have generally been considered as states of cerebral agenesis or simple defect in motor tract development. While cerebral agenesis probably often does define a condition not dissimilar to that resulting in the flaccid diplegias, it by no means is definite enough as a descriptive name to represent the series of pathological events or the nervous mechanism concerned in the formation of the clinical symptoms seen in this disease.

Just as there are varying degrees of spasticity and weakness in the voluntary muscles of the classic type of cerebral palsy of children, so there may be seen, though naturally less often, a combination of spas-

* Read before the Section of Orthopedics, New York Academy of Medicine, Dec. 27, 1912.

1. Förster: *Deutsch. Arch. f. klin. Med.*, 1909-10, xcviii, 216.

ticity and flaccidity in one and the same case. Often side by side in the one limb or segment of a limb flaccid muscles may be found in some otherwise spastic cases, and occasionally one of the lower extremities may be found flaccid and the other spastic, or still more rarely, the upper extremities may be flaccid and the lower spastic. The not infrequent freedom from involvement of the upper extremities while the legs are spastic in a common diplegic is not lost sight of in making this statement. As the clinical material is more carefully analyzed and proved by autopsy study we shall learn to distinguish both the extent and depth of lesion in the palsied brain and possibly arrive at a better prognosis than now obtains.

The flaccid (atonic-astasic) type is even now thought to be an extreme rarity by many. One finds, however, many incomplete descriptions or mention of the condition throughout the extensive literature of cerebral diplegia without the state being diagnosticated as such, and so far as I am aware no one has attempted to explain the lesion underlying the condition or its pathogeny. Förster has called particular attention to its failure of diagnosis and the difficulty of explaining the character and type of the lesion which causes the flaccid syndrome. In 1899, Batten² cited the fact that pediatricians not infrequently encountered cases of ataxy, incoordination, disarthria and hypotonia in children during earliest infancy. The cases which he recorded differed considerably from each other, but he thought he saw a sufficiently marked similarity in all the cases of his group-study to give it the name of cerebellar diplegia. In his type, however, it will be remembered the condition was for the most part transitory, or at least largely removed in the course of a few years. His last case of record, however, remained nearly stationary and without material improvement, and was also attended and followed by nearly complete mutism and imbecility. Its description nearly paralleled those which are the subject of this paper. Inasmuch as Batten's cases never came to autopsy, he had to content himself with postulating the nature and seat of lesion. He held the essential lesion to be a vascular injury to the cerebellum, its peduncles, and sometimes to the mid-brain itself. At that time our present knowledge of more exact functions of the cerebrum and cerebellum, especially their corelationship in disorders of equilibrium and regulation of voluntary movements, had not been attained, and his explanations were not so satisfactory as they might be. The whole subject does not seem to have been pursued further by him or others.

2. Batten: *Trans. Clin. Soc., London*, 1902, xxxv, 205.

TYPE OF CASES EMBRACED IN INFANTILE CEREBRO-CEREBELLAR
DIPLEGIA

The type of case embraced in the flaccid syndrome is very striking. So striking, in fact, that had it not been entirely excluded from the nosological group of palsies of children and classed with agenetic defects, it must not have lacked a more definite study long ago.

The developmental history of the affection is as follows: The child is usually born without asphyxia in a non-instrumental delivery. The intrauterine life of the child in the majority of cases seems to have been uneventful. Only in rare instances has there been good evidence of trauma to the pregnant mother during gestation of the after-affected child. One of my cases was an instance in point. Even under such circumstances many injuries of the like or even more serious character have been of frequent report without having entailed any wide extent of neuraxal lesion as seems to be in evidence in our palsy type here described. The mooted points in its pathogeny will be considered later, in connection with and following the probable pathology.

Usually nothing abnormal in physical and mental make-up is noticed in the child for the first few months after birth. The unusual becomes evident only at that age when the child attempts movements of its own volition, at about 1 year or 18 months of age. It is then noticed for the first time that the child cannot sit up or hold up its head, much less stand. The motility of the arms and legs is normal. The child commonly lies on its back with lower extremities bent considerably at the knees and hips and strongly abducted and rotated outwardly. The legs can be easily extended, and once done, remain so without position. It is then seen that the whole musculature of the body is relaxed. There is no atrophy present, nor does any succeed at a later date. The electrical excitability remains normal. The most striking symptom is the enormously exaggerated mobility of the joints. Up to this point in the description the condition is not so very dissimilar in appearance to a myotonia congenita to which infantile cerebro-cerebellar palsy may be allied in some of its essential features in defect of muscle tonus. However, the analogy ceases with the similiarity in loss of muscular tone. Further examination of infantile cerebro-cerebellar diplegics shows that there is no muscular resistance and the different segments of the limbs can be maintained easily in very arbitrary positions without pain. (See photographs.) The positions in which the limbs of the child can be placed recall those actively assumed by contortionists. This extreme hypotonia has naturally an intimate relationship with the absence of voluntary motion and station. In later years, as the child grows, an ataxy of cerebellar type is superadded and incoordination of all the extremities is marked whenever the child attempts to stand or walk.

Physical examination reveals a wide range of alterations of the reflexes. From increased knee-jerk to a marked diminution or rarely an absence of the same may be noted. As would be expected, ankle clonus is absent while the Babinski reflex may or may not be present. Usually the mental deficit amounts to idiocy or a low degree of imbecility, at least. The mental state undergoes little change with advancing years. There is always a marked speech defect, in the majority of instances amounting to mutism; in others, two or three words only being spoken. While an intrauterine pathology seems likely for this palsy type, not infrequently one finds cases of extrauterine insult either at the time of birth or as the result of a meningo-encephalitis of wide extent. In such cases, though the child may have been able to speak fairly well, a more or less complete loss of speech and idiocy succeeds. As would be expected, the later after birth the brain insult is sustained, the less typical is the clinical picture as above outlined. Still further, one sees other transition cases of flaccid-spastic type which rather insensibly shade out into the classic spastic syndrome, pure and simple. In the transition cases, as not a few of Förster's and one or two of mine herein illustrated show, in addition to spasticity, there are choreic movements somewhat resembling the coarse intention tremor of disseminated disease superadded; even athetosis may be encountered in the impure types. One of Förster's cases had an opisthotonos of the neck and back, whereas complete flaccidity obtained elsewhere in the body. In one of his atypical cases of flaccidity he found a Babinski, distinct spasticity in the lower extremities and increased reflexes. Most of the spasticity in these flaccid cases is developed in looking for resistance.

However, no one should confuse these atypical cases with the classic type of infantile cerebro-cerebellar palsy, the object of our study here. The above digression is made merely to show that under varying conditions of causation very nearly identical nervous structures may be involved in two disorders of the neuraxis essentially differing in pathogeny and anatomical pathology.

The prominent and constant symptoms in all cases, as previously mentioned, are the pronounced flaccidity, the mutism and idiocy. These symptoms establish the disease type. All the other symptoms may vary in special cases, but the symptoms just enumerated are constant. Later, we shall try to make plain the reason for the same.

The positions of the arms and legs are not fixed by proper muscle tone. There is discontinuity of movements, shown in the absence of a cadence of the rhythm and release between the flexors and extensors. A dysmetria, or a lack of proportionate adjustment of the movements to the object in hand, also occurs. If the limbs are left to themselves they follow the action of gravity, and when let fall from a lifted position

behave in as lifeless a manner as a lifeless extremity of a recent hemiplegic. Yet even here the analogy fails of further comparison, for the passive motility of the limbs in infantile cerebro-cerebellar diplegia is far greater than in any hemiplegic, and the extremity offers no resistance to extreme flexion or extension. The knee can be hyperextended and then the heel may be easily carried back to the buttocks or placed on the shoulder of the same side with no expression of discomfort whatever. (See photograph.) The increased passive mobility is equally observable in all joints and in all their movements. Even though one bear in mind the frequent looseness of joints in children, one notes here a distinct pathologic flaccidity in muscles and joints.

As a natural corollary to the flaccid joints and muscles one understands why these children cannot stand. Many of the children can assume no posture at all and always collapse like a jointed doll toy. In some respects the foregoing phenomenon is not unlike the temporary flaccidity not infrequently seen in spastic diplegics after extensive dorsal root section has been undertaken. In the severest grade of the flaccid diplegics the child cannot support the head on the trunk nor hold the trunk erect in the sitting posture. In some mild cases, as the child grows older, one may hold the child by the armpits (see photograph), just permitting the feet to touch the floor. This procedure appears to induce a sort of epileptoid trepidation in the lower extremities, an incoordinative dance. The movements are wild and bizarre like a jumping-jack toy. The effort, be it attempted at volitional standing, or simply a reflex, quickly exhausts the child, fatigue being rapid, when the movements become labored and feeble. After a short period of rest the movements may be reinvoked as before. Rarely in any of these children as they grow older is there an entire absence of voluntary motion of some sort, and in most of them considerable motion is seen in earliest life. Such movements are seen better when the child sits, being held, or better still, when it lies in bed on its back. This statement, however, should not militate against the former statement that in the first few weeks of life these children are, at least in the vast majority, passive and inactive in all movements. This passive state is frequently overlooked as a similar inactivity is not unusually seen in obstetric and pediatric experience where prolonged or difficult labor has resulted in the birth of asphyxiated or puny children. The early observations of the child's inactivity are brought out in sharp contrast by the suddenness with which it begins in the course of a few weeks to show marked and unusual ability to move the limbs strongly. This fact is frequently commented on by the mother of the child, who is much mystified at the further lack of the child's motor development. It is also in sharp contrast to the gradual acquire-

ment of power in spastic diplegic cases. The point is especially called attention to here to show the remarkable effect on even slight development of motility on the part of the growing cerebellum when the cerebellar "brake" is absent or withheld. One must discriminate between purposive movement and those automatically made, as here even in severe cases, while the child is lying recumbent, the head may be moved backward and forward, and exceptionally the head may be jerked about violently, but fixed positions implying purposeful and therefore coordinative movements are never possible. Some of the muscles, on command, may be felt to grow rigid with voluntary effort, but the latter are unproductive in locomotive effect for the other muscles necessary for a voluntary effort refuse to act. This is essentially a disorder of the cerebellar function.

The entire picture of flaccid diplegia or infantile cerebro-cerebellar disease may be summed up as follows:

The defect in active or voluntary motion is due to a complete lack of cooperation on the part of the antagonists and their collateral synergistic muscles, while the immediate flexor or extensor muscles may act with energy. This, too, is an essential defect in cerebellar function.

In looking for a comparison for the flaccidity in this disease, one is strongly reminded of that seen in poliomyelitis, or, better, tabes, though more pronounced in infantile cerebro-cerebellar diplegia. Probably the underlying cause for flaccidity in the two, tabes and our subject, is not so very dissimilar, as we shall see in the theoretic portion of this paper.

The disorder of motility, the wavering tremor, is much like a minor chorea. That the excursion of movement is large is due to two factors, the hypotonia and the dysmetria and unequal control in opposing muscular forces. The condition, as might be expressed, is seen most exquisitely in those receiving their disorder in earliest intrauterine or intrapartum life. The injury and wide destruction of the lesion in cerebral motor function accounts for the persistent disorder, as the anterior cerebrum, the frontal, parietal and temporal lobes can substitute cerebellar functions, and many functions of the cerebrum, as regards motility, reciprocate with cerebellar ones. We must permit in the disorder under study as wide a latitude in clinical expression as that of the ordinary type of Little's syndrome or cerebral diplegia; for in both we are dealing with the negative and positive manifestations of neuraxal symptoms — a diseased nervous system trying to compensate and right itself while in process of development — and especially, too, in that part of nervous functioning, that of motility, acquired in greater part after birth. These are essentially cerebellar functions *par excellence*.

PATHOLOGY AND PATHOGENESIS

What is the connection between these symptoms and an intracranial lesion? Actual proof and study of pathological material we have none. Förster's two autopsied cases are beside the point as they were not classic flaccid cases. The mental defect, idiocy, did not obtain, nor was there complete mutism. They had epileptic convulsions. In passing, I may say that one of mine had convulsions in the first two years of life, but fits have now been absent for two years. One still has fits, and another has trembling fits, a form of generalized cloni as has been noted in recorded cases of cerebellar agenesis. When one searches for similar clinical phenomena in other nervous disease states, one is at once impressed with symptoms recorded in cases of cerebellar disease, particularly the atonic astasic form of cerebellar ataxy, but the history and evolution of the two affections radically differ. For instance, the latter in its pure form shows no extreme extensibility of the muscles and looseness of the joints. There can be no doubt, however, that somewhat similar structures, in part at least, are involved in the two affections.

In closing the clinical portion of this paper on infantile cerebro-cerebellar diplegia, I submit we have to deal here primarily with an intrauterine disorder, which may either be the result of inflammatory or hemorrhagic injury, or, which is probably more plausible, an agenetic disorder of the cerebellum and a wide and extensive defect of the fore-brain, especially the frontal, parietal and possibly the temporal lobes of the cerebrum, as a result of which hypotonia, dysmetria, ataxy, incoordination, mutism and idiocy result.

Let us review briefly the more recent experimental and clinical evidence of cerebro-cerebellar symptoms to make clearer the pathologic anatomy of our disorder of the infantile cerebro-cerebellar diplegic.

THEORETIC CONSIDERATIONS OF THE RELATIONSHIP BETWEEN CEREBELLAR
FUNCTIONS AND THE FOREBRAIN, AND THE BEARING WHICH
THESE HAVE ON THE CEREBRO-CEREBELLAR PALSY
OF CHILDREN

While it is generally conceded that the cerebellum plays little or no part in the organic functions of the body, of sensibility or intelligence, there are few functions in life concerning the relation of movements in which the cerebellum does not enter. As regards the mildest grade of intelligence defects occasionally noted in cerebellar disease, though the case be a specific and limited lesion of the cerebellum solely, the intellectual defect would be understandable by finding a certain mental deficit whenever there is a lack of mechanical adaptation of bodily movements from which the psyche receives no small impetus to its proper development; the individual subject might be so engrossed in minute

and automatic adaptations of bodily movements in cerebellar disease as to narrow the limits of the actual field of experience from which the psyche gains its greatest incentive for growth.

In considering the relationship of the cerebellum to motility, it must be said that some investigators still believe that this organ is the center of energy, or reinforcement of muscle tonus, though the majority of writers seem to entertain the idea that it is purely a center of coordination of all bodily movements. Still others limit its rôle to that of equilibration only. It is fairly well known that the peripheral excitations are gathered together in the cerebellum on their way to the cerebral cortex. It is therefore conceivable that the cerebellum may be an active organ in elaborating or controlling muscle sense; at least to that point of making it a relay station for these incoming excitations and thus it would necessarily play a rôle in the variations of muscle tension so essential for proper equilibration.

As a matter of anatomic moment, we know that the gray substance of the cerebellum and pons has a crossed function, while the nuclei of the latter communicate directly with the cerebral cortex, especially the forebrain of the same side. This association is made principally by the frontal bridge tract through the crus cerebri to the frontal, the ascending parietal lobes and the paracentral lobule. A portion of this tract known as "Türk's bundle" takes its specific origin from the middle segment of the second and third temporal convolutions exclusively. Therefore we see that there is a close functional relationship between the cerebellar and the cerebral cortex. The cerebral zones here considered, which are projected on the cerebellar cortex, are primarily sensori-motor zones. The second and third convolutions, however, are ordinarily considered as centers of impressions of labyrinthian origin which pass from the middle cerebellar peduncles. The interrelationship of certain portions of the cerebrum and cerebellum is proven in part at least by comparative embryological studies which always show a parallel between the development of the pons, the median cerebellar peduncle and the cerebral cortex.

Even though both the hemispheres of the cerebellum are destroyed in animals, it is known that reeducation takes place in time by means of the reciprocating functional powers of the cerebrum. In cases of cerebellar agenesis, coinciding with a cerebral defect of the forebrain, mutism is almost invariably found in both experimental and clinical studies; dogs do not bark and children do not speak after lesions of these parts. The reflexes in total destruction of the cerebellum are increased, the "brake" on the reflex arc is removed. However, whenever a wide destructive lesion of the cerebrum is entertained, especially in the forebrain, together with cerebellar ablation, the knee jerks are markedly diminished or lost. This variation in reflexes under experimental conditions explains

much of the wide variation seen in the reflexes of the clinical types which we have under study. Moreover, when both middle cerebellar peduncles are excised in animals, they lose power to stand or walk.

In applying experimental evidence on the cerebellar functions to explain our clinical type, we must proceed with due caution, as the symptoms of hemorrhage and softening in a fully evolved neuraxis are quite different from those of lesions of a slow and progressive disorder, and, moreover, both groups of symptoms are still more variant when they occur at birth, in the cerebellar organ, which is still outside the realm of every-day use in the new-born child.

Many of the cerebellar experiments cited in literature are incomplete, and in others the lesions are not completely confined to the limits of the

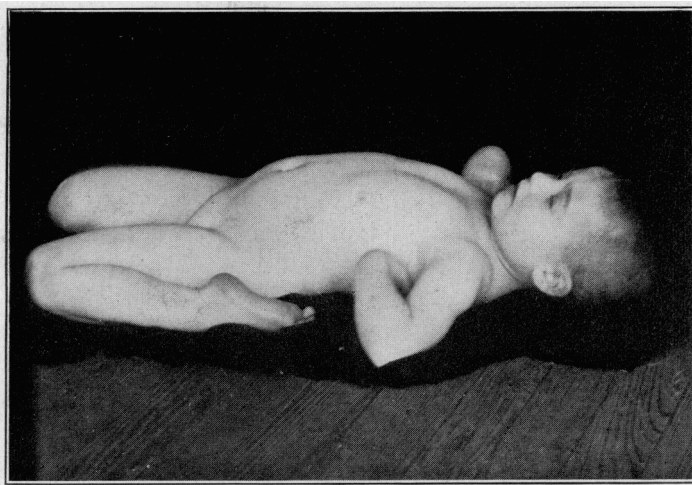


Fig. 1.—T. S., 5 years old. At 1 year of age had fever and convulsions; five weeks sick: lost all speech and slight ability to stand. Now all extremities are very flaccid and hypotonic, greatest in legs, right arm and neck. Bilateral Babinski, knee-jerks +. No ankle clonus. Coarse incoordinate movements in arms, legs, head and trunk.

cerebellum alone. As is known, the symptoms following cerebellar disease are staggering gait, widening of the base of support and a disturbance of equilibration. The patient falls to the side or backward, according to whether one cerebellar hemisphere or both are equally involved. To this picture may be superadded an intention tremor or cloni in the upper extremities and certain incoordinative and ataxic movements in the lower limbs and rapid fatiguability. Vertigo and various ocular disturbances as well as nystagmus are less common. While most authorities agree that a real paralysis, such as that of ordinary hemiplegia in the adult, cannot be produced as a result of disease of the

cerebellum or experimental ablation of the same, most writers are agreed that a certain type of motor defect which *amounts* to paralysis may be seen. The defect, too, is seen in *all* muscles in contra-distinction to that seen in ordinary hemiplegia, where special groups of muscles, such as a paralysis of dorsal flexors, flexors of leg, extensors of hand and external rotators of the arm are chiefly if not exclusively affected. Further, in the motor disability of cerebellar disease there is always an absence of

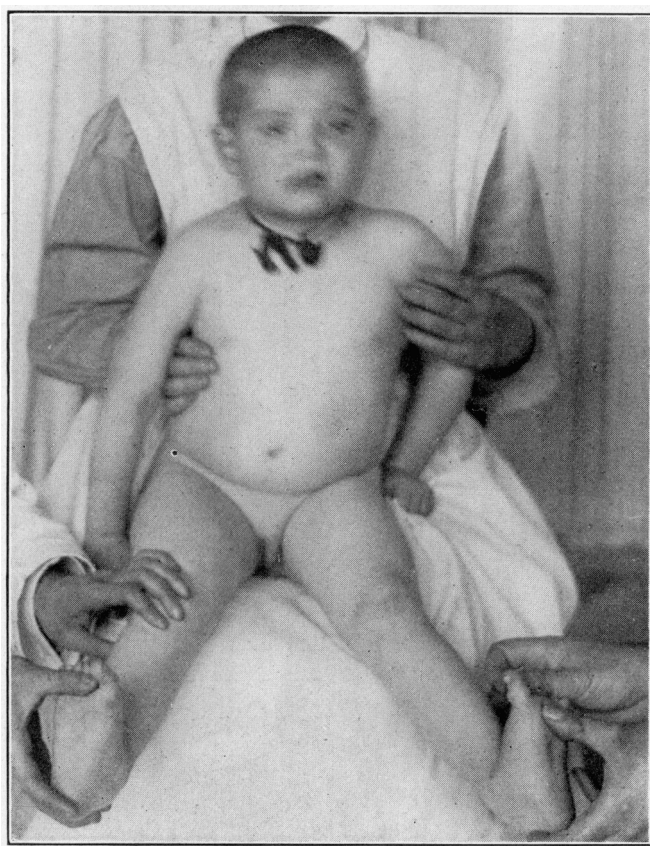


Fig. 2.—T. S. Showing flaccid ankle-joints.

contractures. Finally, the reflexes are exaggerated *without the occurrence of spasticity*, and in the pure uncomplicated type of cerebellar injury or disease there occurs no epileptic trepidation nor Babinski. The foregoing facts all point to a better understanding of the anatomico-pathological condition which has been postulated for our type of infantile cerebro-cerebellar diplegia.

In reviewing the clinical reports on cerebellar disease, one is impressed with the entire absence of symptoms in some cases even though a complete cerebellar agenesis has occurred. Cambetti's case is an instance in point; the lesion was clean cut, showing an entire absence of the cerebellum without any other defect. The child developed slowly, was backward in all essential organic movements and spoke with diffi-

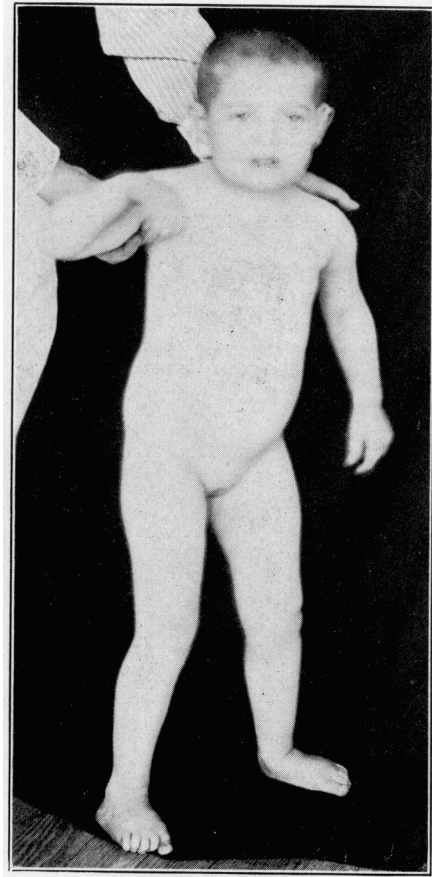


Fig. 3.—T. S. Wide base of support. Cannot stand alone or walk.

culty and hesitation. In Anton's case, that of a little girl of 6, all her movements were slow and incomplete. She sat up, stood and walked only after 4 years of age. Even then she was unable to walk freely; she moved about only by artificial support. Incoordination, staggering and frequent falls persisted. Rapid fatiguability was very marked. The speech was not very clear, but no tremor was in evidence, and the kneejerks were exaggerated. In one of Shuttleworth's cases there was gen-

eral muscular weakness, tremor of the hands and arms, and the child remained an imbecile. Borell's case showed an extraordinary bending of the legs in walking (due to looseness of joints). In all cases there have been motor disturbances and considerable hypotonia. In all, there has been an absence of the rhythm or cadence of normal movements. The peculiar defect in attempts at walking in cerebellar patients is quite characteristic. The foot is frequently advanced ahead of the trunk, while the latter does not follow up the attempt at progression by the leg, consequently there results, as in my cases, a sort of paddling movement of the feet without trunk advancement which is necessary for normal bodily progression.

From a careful search in the literature, one is impressed with the fact that *bona fide* cases of primitive atrophy of the cerebellum, which

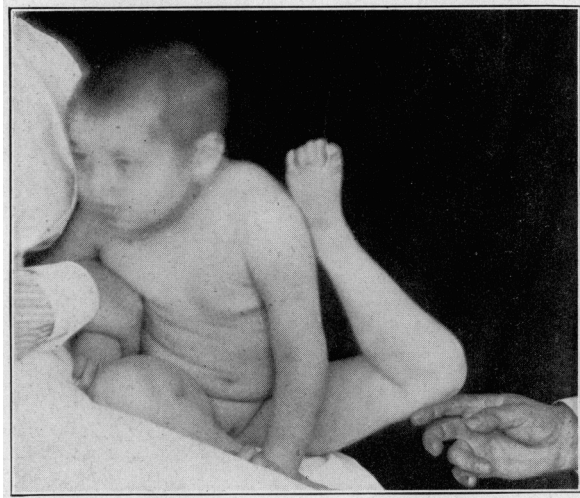


Fig. 4.—T. S. Showing hypotonia of knee-joints.

would be most pertinent to our study, are very rare, and if one includes those cases in which other parts of the neuraxis are absolutely healthy, the case material is exceptional. There can be little doubt that a *lack of proportion in volitional movements*, seen in nearly all cases of infantile cerebro-cerebellar diplegia, a phenomenon named dysmetria by Thomas and Babinski, is a sure sign that the cerebellar portion of the neuraxis has been seriously crippled in this palsy. If one examines these children carefully he finds the essential defect in volitional movements is due to a contraction of the muscles in several stages and not in a tonic fashion as normal muscles are; therefore, a sort of cloni occurs which may amount at times almost to an epileptic crisis of cloni, as noted in one of my group. This speaks for a serious damage to the coordinating func-

tion of muscle rhythm which is presided over by the cerebellum. The rapid fatiguability well shown in my material is to be accounted for, not on the basis of a weakness or a paralysis, but therein the automatic effort of repeated testing out on the child's part of large groups of muscles calls for a great expenditure of energy and rapid fatigue results.

It has been found that substitution of the function of one lobe of the cerebellum by the other occurs up to a certain point, while in the absence of both hemispheres, as previously mentioned, the frontal lobes contribute an important help in maintaining equilibrium. An interesting case which was proofed by autopsy and found to be olivo-fronto-cerebellar

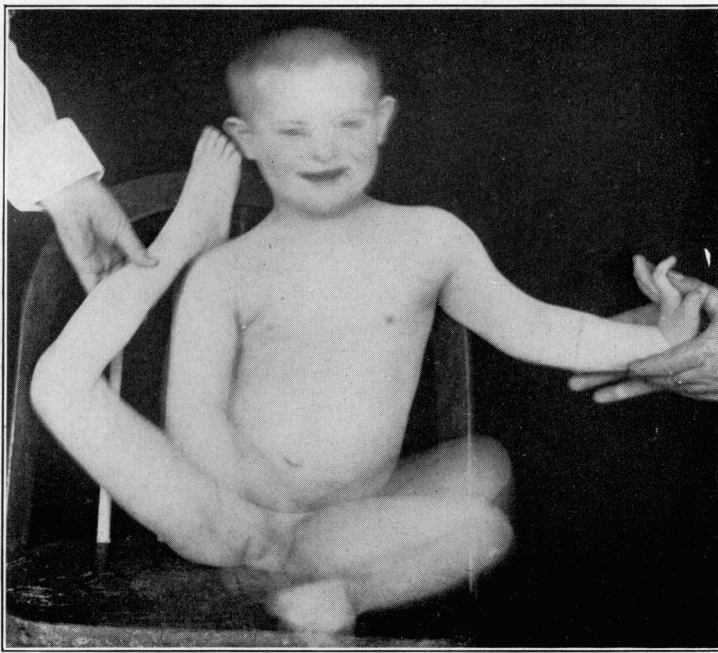


Fig. 5.—F. N., 8 years old; no history of palsy or injury. Never talked nor walked. Classic flaccid hypotonia of muscles and joints. Some ability to stand supported, but very marked incoordination; most pronounced in right leg. Wildly paddles legs when walking is insisted on. Apparently understands two or three simple commands. Idiocy. No Babinski, and knee-jerks —. No ankle clonus.

atrophy, showed disorder of gait and station, nystagmus and scanning speech; but there was here also a bilateral atrophy of the cerebellar peduncles, while the pyramids remained absolutely intact. The patient was therefore not really paralyzed, and there was no real motor weakness. All this goes to prove that any disorders of movement in atrophy of the cerebellum are accounted for by interruption of the fronto-bridge tract, and, secondly, if the latter remains intact the cerebral cortex may sub-

stitute in a large measure the defective functioning of the cerebellum in regulating muscle movements. The substitution is more easily made, and the more complete, the earlier it takes place in the development of the neuraxis, before the nerve centers have been affected by subsequent perverse habits. The individual differences in range and degree of the lesion explain the difference in the degree of recovery in our cases. It is a common fact that whenever there is very wide area of the motor zone diseased dating from infancy, the opposite cerebellar lobe is atrophic as well as the pons of the opposite side, and the olive of the same side as that of the cerebral lesion. This is very well shown in a case of my own reported some years ago in an epileptic girl. In such case the atrophy of the cerebellar cortex, the middle cerebellar peduncle and degeneration

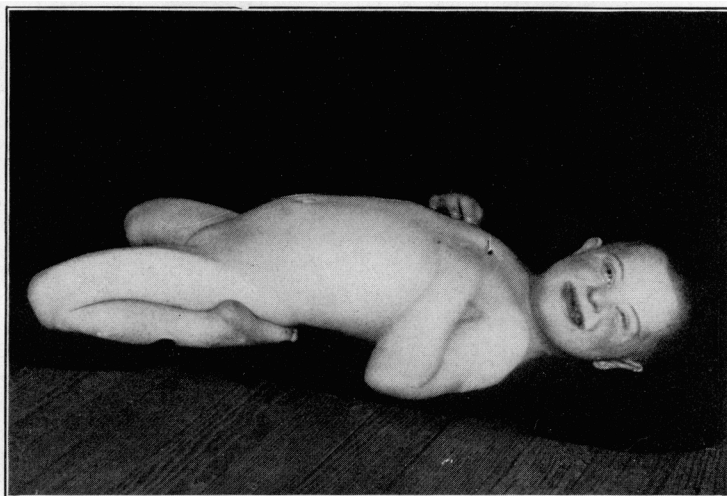


Fig. 6.—F. N. Shows hypotonia of legs of same child, who is easily placed in an attitude which would be very uncomfortable to a normal one.

of the peduncular path followed into the gray substance of the pons, the pontine nuclei. As a system, this fasciculus which had degenerated is known as the cerebro- or fronto-cerebello-pontine tract. The cerebral cortex of the frontal, parietal and temporal regions is projected on the lateral lobes of the cerebellum of the opposite side and also to a slight degree on the lateral lobe of the same side by the intermediation of the cerebral crus and the pontine nuclei. A consideration of the spinal medullary centers which are projected on the vermis proves that to some degree the functions of the vermis and the lateral lobes are different. It is the first system which suffers a defect in the infantile cerebro-cerebellar diplegias. This system, once disordered, as in infantile cerebro-cerebellar diplegias, would bring about a defect in equilibration primarily.

Finally, we may summarize our subject as follows: We have here under consideration an extreme defect disease of the cerebro-cerebellar function with relationship to bodily movements, of an unknown pathogenesis (probably based largely if not solely on a developmental defect) which is for the most part intrauterine in origin and which we find expressed in clinical manifestations of atonia, astasia, hypotonia, mutism and idiocy. We believe it constitutes a distinct type of palsy of infancy. The disorder not only affects isolated movements, but particularly the *association* of movements or the motor synergies so-called. The movements by themselves as shown in the type are not incoordinated as in tabetics, but are characterized by dysmetria and discontinuity. When-

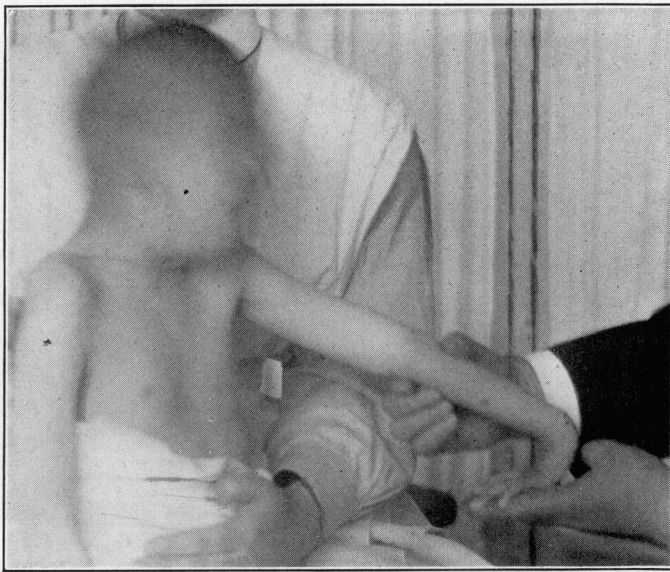


Fig. 7.—M. D. Showing hypotonia of wrist.

ever the child's movements relate to the maintenance of an attitude, especially in sitting or standing, there is instability, wavering from side to side, or astasia. The children show great disturbance of the reactions to equilibration, due to the fact that the defective cerebellum fails to effect and hasten the reestablishment of equilibrium, much the same way as it fails to make isolated movements precise and regular. Again, the defective cerebellum fails to apportion the measure and the continuity of movement. There is instability and imperfect reactions to equilibration by a special tonic action. This latter function is co-governed by the cerebrum and to some degree by peripheral stimulation. The defect in cerebellar function shown in hypotonia is not compensated for by a dis-

eased cerebrum. The latter organ being unable to substitute or take up the cerebellar function there results a persistent dominance of hypotonus throughout the life-picture of this type of palsy. Again, as exquisitely shown in our clinical type, the suppression of the cerebellar functions in the condition or disease under study does not produce a palsy *per se* as ordinarily understood. Motility of the limb remains. It is never

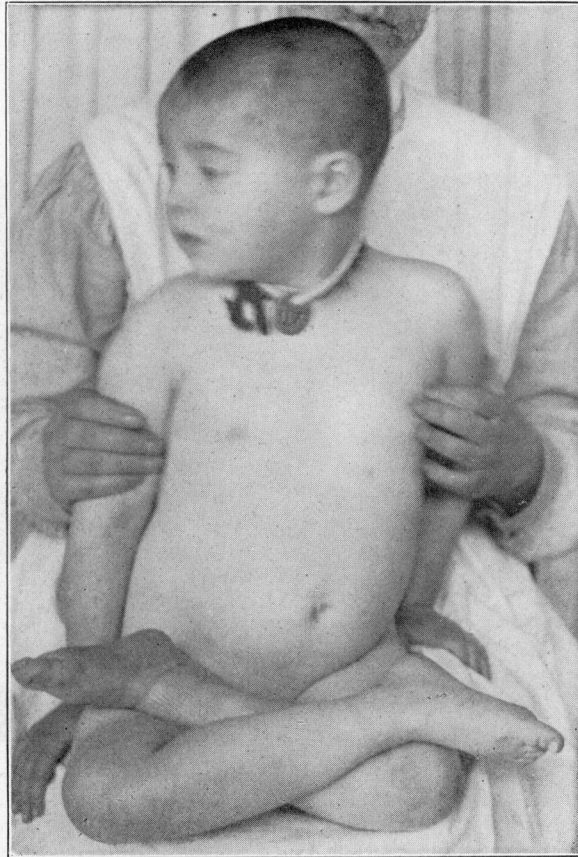


Fig. 8.—M. D. Hypotonia shown in child sitting in extreme “tailor fashion.”

abolished entirely, as some movements or attempts at equilibration probably remain because some degree of substitutional power of the cerebrum is still possible; the greater part of its loss, however, is largely in evidence in this unique disease.

SUMMARY

We may here briefly summarize the points brought out in the foregoing paper. (1) Infantile cerebro-cerebellar diplegia of the flaccid, atonic-astasic type is a relatively rare disorder, which has largely an

unknown intrauterine causation, but which results in a permanent disablement of the motor functions of the cerebellum and forebrain, resulting in (2) a state of mutism and idiocy accompanied by atonia, astasia, dysmetria, incoordination and a generalized palsy in effect.

ADDENDUM

While the data on cerebellar disease is already extensive, that which has a more or less distinct bearing on our type of associated palsy of brain and cerebellum is not so great but that we may be permitted to give a few abstracted case-histories which illustrate some of the special points. It is hoped that citations of this kind may signally assist the

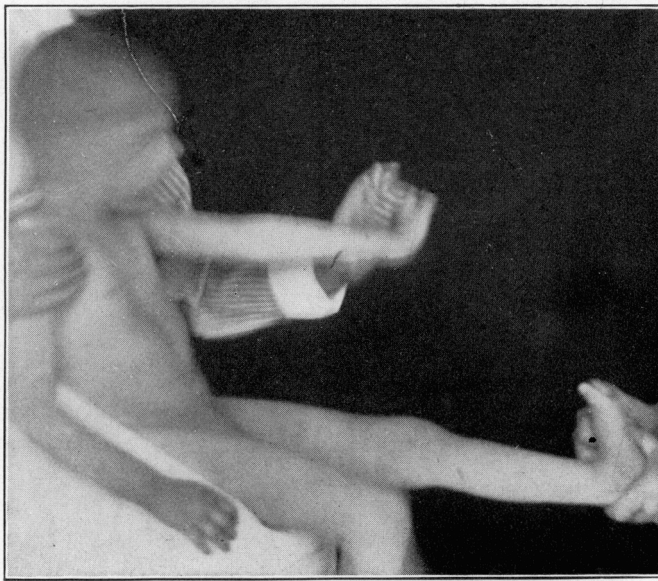


Fig. 9.—M. D., 4 years old; youngest in family of five. All others healthy. Patient born normally at term. Normal mental and motor development (?) until 5 months' old, when, after a few days of fever, diarrhea and convulsions all extremities became limp. Fits continued for two years, severe and frequent. When mother was three months pregnant with patient she had a fire fright and fell downstairs. Child has never spoken, and is a profound idiot. Never walks but often tries unsuccessfully to stand. Cries continually. Bilateral Babinski, vasomotor stasis in hands and feet marked. Knee-jerks + and no clonus. Lower extremities flaccid, hypotonic in muscles and joints. Marked incoordination.

inquirer to orient himself, as it were, regarding the relative clinical and pathological position which the new type of infantile cerebro-cerebellar palsy should occupy in the brain palsies of infancy.

In looking over the monumental work of Freud, one is impressed that at the time his special work on infantile cerebral palsies was written,

some twenty years ago, the ordinary conception of a flaccid palsy was indicative of an abortive case, and in such afflicted individuals one might expect them to recover (like Batten's cerebellar diplegia of 1898-9). Freud regarded the existence of a permanent flaccid palsy as improbable, as a spastic or rigid state *might* have existed at some period unobserved.

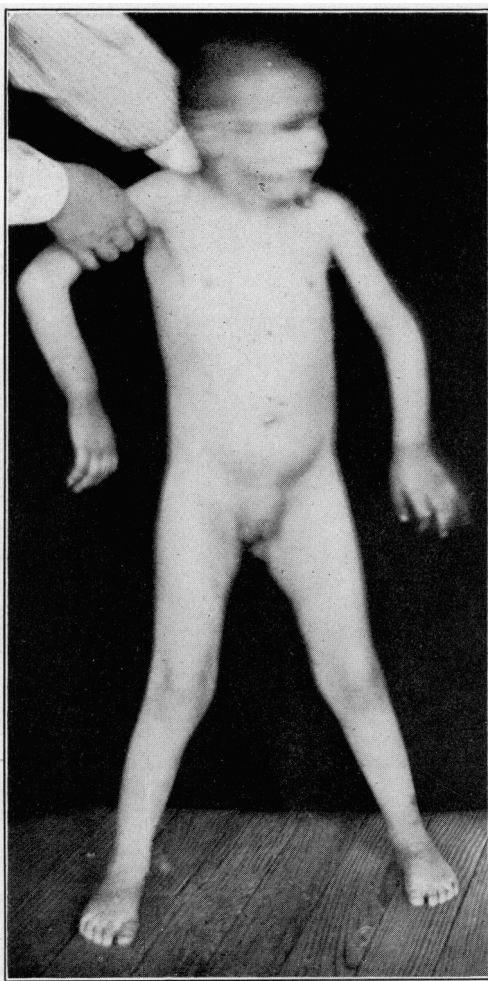


Fig. 10.—M. D. Shows typical straddling attitude of child unable to walk or stand alone. Note blur in photograph from incoordinate movements of head and left hand. Back and head supported.

In Adler's 134 autopsies in cerebellar lesions no cases occurred in the newly-born, or even in the first two years of life, and very few in children of any age.

That cerebellar agenesis or atrophy *per se* cannot cause a flaccid state is shown by the fact that some individuals born with a rudimentary cerebellum seem to show no motor disorders. On the contrary, if the cerebellar atrophy is secondary to a cerebral lesion, or if the cerebellum

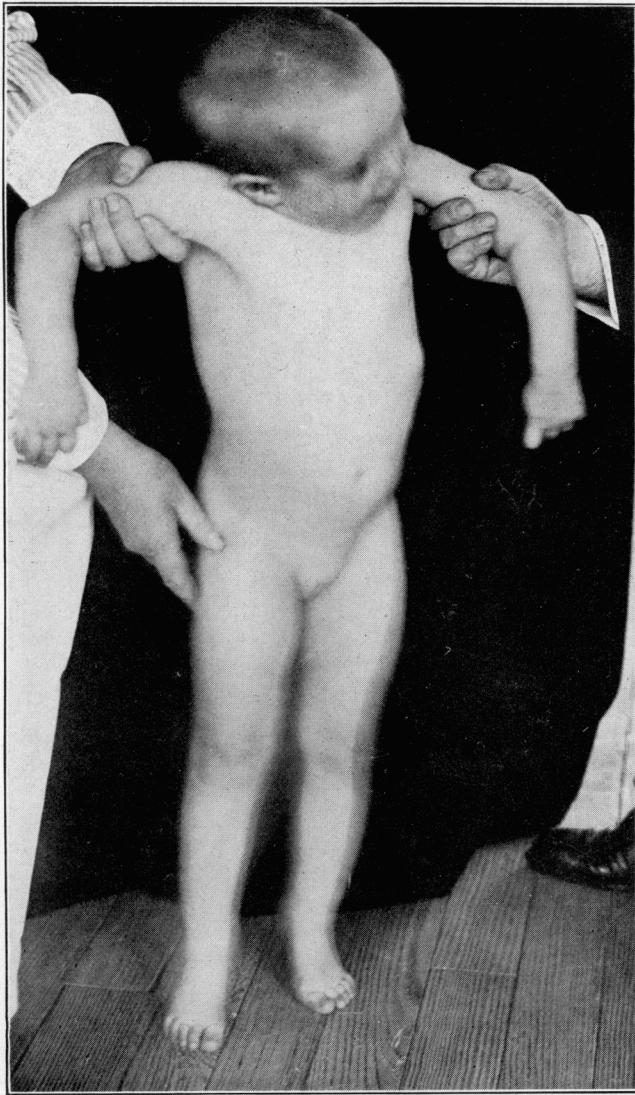


Fig. 11.—L. S., 4 years old; idiot; cannot talk, walk nor stand. Flaccid and hypotonic in all extremities. Marked incoordination. "Dancing doll" type when held standing. Note inability to hold head erect. Knee-jerks +. Bilateral Babinski. No ankle clonus.

and cerebrum are both agenetic, very complex symptoms present themselves. A typical illustration of each will be given.

The first case is one of congenital absence of the right hemisphere, reported by A. Pearce Gould.³ The man lived to be over 80 years of age, and had worked for sixty years for one family as a gardener. Nothing

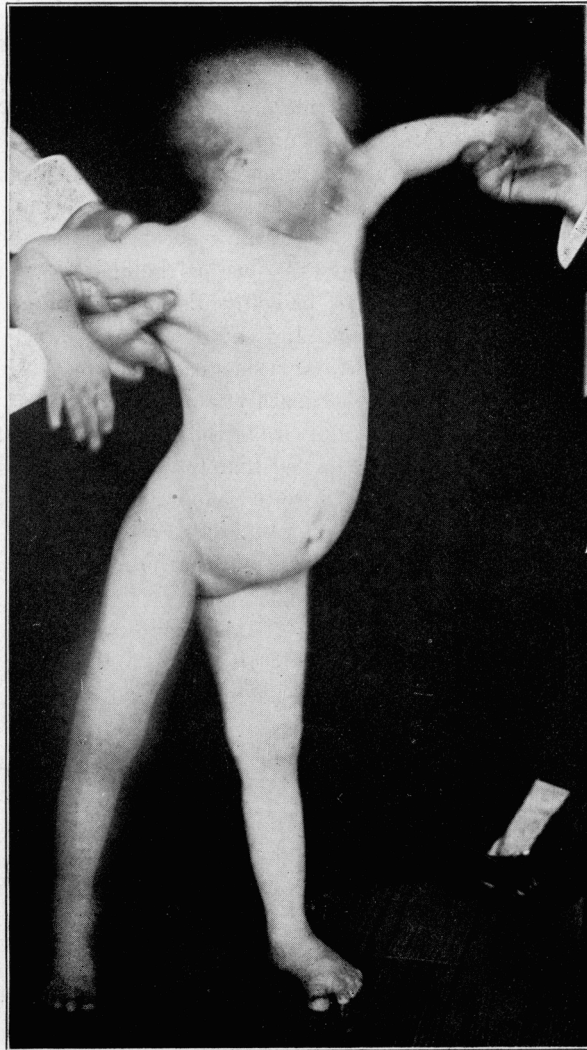


Fig. 12.—L. S. Shows result of an attempt to get the child to walk. Head is in incoordinate associations. Legs held wide apart, the right one bearing little weight is in dancing vibratory motion.

else was known of him, as shortly before death he had been moved to an asylum where he was unknown. The left half of the cerebellum was

3. Gould, A. Pearce: *Trans. Pathol. Soc., London*, 1881, xxxiii, 6.

normal, and there were no evidences of disease in the vicinity. That the condition represented an agenesis and was not the result of some ancient lesion well appeared from the aberrant course of the lateral sinus, which occupied much of the cerebellar fossa; the flattening of the bone externally, the negative evidence as to tumors, abscesses, etc., and finally the normal quality of a rudiment of the left hemisphere. This agenesis had led to no serious motor disorder.

In explanation of the lack of motor defect, it may be fairly urged that this case but illustrates the point made in the theoretic portion of this article; one sound lobe of the cerebellum may compensate for even an entire absence of the other lobe. This case with retention of normal motility is quite similar in all essential respects to that reported by Hitzig.⁴ The patient, female, was first seen at the age of 32 years. The mother stated that she had never had any disturbance of motility, had learned to walk at the proper age, and could leap and dance. She was backward in school studies, but married in later life. Nine months before consultation she developed paralytic dementia from which she died. The author shows elaborately that the case was one of agenesis, not of secondary atrophy. He believed that the cerebrum probably acted vicariously in these cases. The two cited cases bear out Luciani's⁵ contention that agenesis of one-half of the cerebellum causes no symptoms, while in agenesis of the whole organ incoordination and hypotonia surely follow.

In one-half the cases recorded in literature one finds bilateral incomplete agenesis in a latent form and in the other half ataxia, astasia in lower limbs, and frequently in the arms and hands also. Luciani expressly states that the latent agenesis or defects dating from embryonal life the cerebrum compensates for the cerebellar defect, although he throws no new clinical light on our type of palsy in the newly-born. A very remarkable case showing how the other portions of the neuraxis may show compensatory hyperplasia in bilateral defect of the cerebellum is reported in clinical and autopsy detail by Anton.⁶ As it is one of the most exhaustively reported cases on record of its sort, I will give it in brief abstract.

Mother of child delicate; father tuberculous; gestation normal. Labor easy (infant very small) and finished in an hour. Infant nursed normally and later was fed with a spoon. It was noted very early that swallowing was awkward, causing coughing spells. The movements of the limbs and trunks were learned slowly and imperfectly. The child was in her fourth year when she learned to sit and stand, and she clung

4. Hitzig: *Arch. f. Psychiat.*, 1884, xv, 266.

5. Luciani: *Ergeb. d. Physiol.*, 1904, iii, 1, 259.

6. Anton: *Wein. klin. Wchnschr.*, 1903, xvi, 1349.

to objects for support. She could never walk backwards, and all movements were weak. She became quickly exhausted after efforts of any sort. Speech was defective, low and infrequent. She was somewhat apathetic and greatly irritated by certain sounds. There was no tremor of the hands, but they became exhausted so quickly that objects were passed from one hand to the other and back again. Attention was difficult to fix. At the age of 4½ years she died of acute laryngitis. A full autopsy report is given. The absence of the cerebellum was thought to be due to porencephalic softening.

The parts which showed the compensatory over-development were the pyramidal tracts, the nuclei of the posterior columns, the gelatinous substance of the trigeminus with its sensory nucleus and locus ceruleus. The thalamus and corpus striatum were well developed and showed no sign of degeneration. The cerebral cortex was thicker than normal—a relative hyperplasia.

Ewald, Thomas and others have proved that compensation of this sort does occur. It is, of course, only partial, but may improve progressively.

From the above brief citations from the literature of our subject one is impressed that the whole subject is entirely covered experimentally and pathologically, and it proves that (1) unilateral agenesis is compensated for by the remaining intact lobe, and (2) that bilateral agenesis may be compensated for by the cerebrum. Evidence is not yet forthcoming to show in cases (like ours) in which agenetic defect of both cerebellum and forebrain is postulated to exist, in what way the multiple lesion is induced, whether the cerebellum or the cerebrum suffers the initial damage or whether both are simultaneously injured, and how. The pathogenesis of the disorder still rests in the darkness and mystery of intrauterine pathology.

In reviewing the literature of cerebellar disease, I wish to give proper credit to the excellent work and résumé of Dr. André-Thomas⁷ of Paris. I have consulted the original articles on cerebellar research and have found his exposition so complete and satisfactory for the purposes of this article that I have borrowed the substance of many of his conclusions of the co-functioning of brain and cerebellum where such have been particularly pertinent to prove the hypothesis of my thesis. All the statements in this paper based on the morbid anatomy and physiology of different workers on cerebellar disease have received the critical sanction of Thomas, as above noted.⁸

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7. André-Thomas: "Cerebellar Functions," *Nerv. and Ment. Dis. Monograph No. 12.* Jour. Nerv. and Ment. Dis., New York, 1912.

8. The clinical material shown in the photographs illustrating this article is drawn from my service in the Randall's Island institutions for paralytics and mental defectives.