

On athetosis, or imbecility with ataxia / by T. Clay [sic] Shaw.

Contributors

Shaw, Thomas Claye, 1840-1926.
Royal College of Surgeons of England

Publication/Creation

[London] : [publisher not identified], [1873]

Persistent URL

<https://wellcomecollection.org/works/g6whzvXu>

Provider

Royal College of Surgeons

License and attribution

This material has been provided by This material has been provided by The Royal College of Surgeons of England. The original may be consulted at The Royal College of Surgeons of England. where the originals may be consulted. This work has been identified as being free of known restrictions under copyright law, including all related and neighbouring rights and is being made available under the Creative Commons, Public Domain Mark.

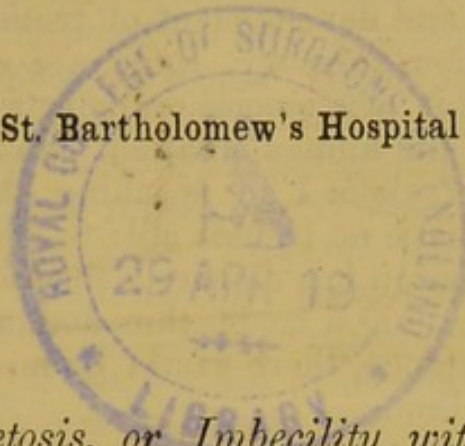
You can copy, modify, distribute and perform the work, even for commercial purposes, without asking permission.



Wellcome Collection
183 Euston Road
London NW1 2BE UK
T +44 (0)20 7611 8722
E library@wellcomecollection.org
<https://wellcomecollection.org>

8.

[Reprinted from St. Bartholomew's Hospital Reports, Vol. IX.] 1873



On Athetosis, or Imbecility with Ataxia.

By T. CLAY SHAW, M.D.

THE term 'Athetosis' has been applied to a peculiar motor affection first described by Dr. Hammond of New York, and since his description alluded to by Professor Gairdner. These two writers refer to a disease attacking persons who, up to a certain age (more or less advanced in particular instances), were in good health, with perfect command over all their muscles. Before reading Dr. Hammond's description, I had noticed a peculiar but limited class of imbecile persons who resemble each other closely in features, bodily development, and peculiarity of action in certain muscular groups. These imbeciles (including persons of both sexes) represent a congenital condition, or at any rate one supervening soon after birth; and they so closely resemble those whose motor affection comes on at a later period, that one is forced to conclude that the central lesion is the same in both sets of cases, and that the history of the one set may explain that of the other. The rarity of the symptoms is shown by Dr. Hammond having only six cases to report, and my experience is limited to about the same number.

The affection may be at once described as an inability to retain a fixed position, owing to a slow but constant action of certain extensor and flexor muscles.

As in Dr. Hammond's instances of secondary disease, so in mine of a congenital affection, it is the muscular inco-ordination that renders them incapable of being of much service to themselves or others. The term 'Imbecility' is difficult of

exact definition, and the class of which I am now speaking is certainly very different from the ordinary imbeciles and idiots met with in asylums, about whose condition of 'non compos' there can be no doubt. Thus, in shape and size of head and ears, arrangement and development of teeth, height of palate, power of articulation, memory, some are of first-rate order, whilst others exhibit deficiencies or defects condemning them to a lower stage of the human creation. So intelligent, indeed, are some of those of whom I am speaking, that were it not for the fact that they have no friends able to look after them, and that their motor lesion has deprived them of the opportunity of learning and pursuing a trade, thus making them dependent upon external assistance, there would be no reason for detaining them in an asylum.

Just as some are rendered idiotic by 'deprivation,' from obscuring of one or more of the special senses, leaving but small means for objective stimulus to the brain, so are these, who otherwise show much intelligence, prevented from developing to the extent they might otherwise have done, owing to the impediment that this constant muscular movement has placed in their way. Continuous rhythmic movement being, then, the characteristic feature, it is as well to describe first of all of what it consists.

Dr. Hammond says: 'There are several diseases one of the chief manifestations of which is involuntary movement, the consciousness of the patient remaining intact. *Ἀθητός* signifies inability to keep the fingers and toes from continual action. Not disorderly, like hysteria and chorea; not tremulous, like paralysis, agitans, and various forms of sclerosis. They are regular, and to some extent under the control of the will. They continue during sleep (?). There is pain in the affected parts; the sensibility of skin is lessened, and a feeling of numbness prevails over the side of the body corresponding to the disordered motility. Of the six cases known to me, two have been women.'

Dr. Gairdner's description is good, and runs as follows:— 'Sometimes the wrist is strongly flexed and at other times it is strongly extended, and very often the fingers are in precisely the opposite position to the wrist; sometimes individual fingers are flexed while others are extended, and all more or less rigid. Under favourable circumstances he can perhaps control every muscle of his arm; but when the rigidity or spasm comes on, particular groups are, for the time being, wholly withdrawn from the will, or nearly so. They scarcely ever cease except

in sleep. By using a certain amount of force you can always overcome the spasm in opening successively the fingers of the closed palm, but an attempt to overcome the spasm of one group of muscles is very apt to set it off in the opposite direction. It seems like incontrollable muscular impulse, without paralysis and without permanent spasm or rigidity, a quasi rhythmic spasm, somewhat resembling the peristaltic movement of the involuntary muscles in the alternating or successive affection of groups of muscles.' Neither here, however, nor in Dr. Hammond's cases, is any mention made of movements in the muscles of the head, neck, or face, which in my instances are well-marked, and resemble in character those of the forearm and fingers. They consist of a slow protrusion of the head forwards and upwards, to one side or the other, and of its retraction downwards and to the other side. As might be expected, the muscles in the sub-occipital triangle are hypertrophied like those in the arms. The facial movements are very extraordinary, and give rise to varying expressions, the most frequent being that of a broad grin, owing to spasms of the retractors of the angles of the mouth and the 'laughing muscles,' the zygomatics, and the lower fibres of the orbiculares palpebrarum, which raise the lower eyelid slightly; the forehead, too, is wrinkled. To this succeeds—or may succeed—a comparatively blank look, from relaxation of these muscles, but the marks of the wave are shown in the furrows left, giving an appearance of age greater than is actually the case. Not that an expression is developed suddenly, for it often happens that one emotion appears to be merged into another, owing to the vermicular manner in which the wave progresses; thus the occipito-frontalis may contract, elevating the eyebrows and giving the expression of wonder, which expression may directly afterwards pass into that of a broad grin, by the contraction of the retractors of the mouth and lower jaw, displaying the interior of the mouth and the teeth. As the corrugator supercilii, or grief-muscle, may also be excited by the wave passing over the face, the expression caused by it mixes at times curiously with the others. It does not appear that the emotion corresponding to the contraction of a certain group of muscles, i.e. to an expression, is excited, for the movement is very transient; and besides, it by no means follows (as some have on the contrary asserted) that the action of certain combined groups of face-muscles evokes the emotion which, as a rule, in the anatomy of expression, precedes the contraction. Persons looking at an actor whose features are strongly worked may assume a

similar expression by so-called sympathy, i.e. sensori-motor action, but the emotions in these persons are by no means at all times the same. A similar state of the expression of an emotion without the existence of the emotion may be often seen in those affected with 'general paralysis of the insane,' where an appearance of grief is by no means uncommon, no grief being really felt, but the grief-muscles being put in action to assist some that are paralyzed. Like the face-muscles, those of the fingers seem independent one of the other, for not only do they bend and extend themselves capriciously, out of unison with the others, but the flexions happen now at the second, then at the proximal joint, while the fore-arm is ever on the move from pronation to supination and back again. The upper-arms are not affected to the same degree as the fore-arms and hands, and only in some are the lower extremities affected in the particular manner. I say in 'some' because, though all are impaired in gait to some extent, yet two distinct kinds of alteration may be discovered; in the one the gait resembles the 'tabic' form so often seen in the 'general paralytics;' and in these persons muscular movements of the toes resembling those of the fingers may be seen; in the other class the defect seems to be a want of co-ordinating power combined with a distortion of the pelvis, owing to spinal curvature, which is more or less pronounced in all the cases, and consists generally of a primary curve in the dorsal region, with secondary curves in the cervical and lumbar.

The following is an analysis of the cases:—

CASE I.

H. J., female, *æt.* 43. Not epileptic; can talk, read, and do needlework. Always been in present condition. Head continually being turned to left side and slightly upwards, through spasm of the right sterno-mastoid and the muscles in the right side of the neck. As to the face, the risorius, zygomatics, orbiculares, occipito-frontalis, are in constant motion, waves passing over them but not causing the least pain. The tongue is slowly protruded from the mouth and withdrawn; and as all these spasms are intensified when she attempts to speak, the articulation is peculiar, the mouth and tongue being dragged in different conditions under the influence of non-co-ordinated muscles. The face is much and deeply wrinkled. The right shoulder is rather more raised than the left, owing to a slight spinal curvature, and both upper-arms are held

DESCRIPTION OF PLATES.

FIGS. 1, 1*a*, 1*b* represent H. J. in different attitudes, following each other very rapidly owing to the wave of muscular spasm. In 1 the action of the retractors of the mouth is well seen, and in 1*a* the second, third, and fourth fingers are flexed, the index being extended. The head is seen to be pushed forward in 1*a* and 1*b*, and turned to the right or left side.

FIGS. 2 and 2*a* represent H. H. taken as rapidly as two consecutive plates could be brought to bear. The change in expression, and the alteration in the position of the head, with pronation of the arm and closing of the hand, is well seen.

FIG. 3 is to show the position assumed by the fingers, the forefinger and thumb of the right hand being extended for the moment, most of the others closed. Both in personal appearance and in movements this man closely resembles the woman depicted at 1*a* and 1*b*.

FIG. 4. L. C. The difference in the facial expression is very marked, and took place very rapidly. The right hand, which in fig. 4 is clenched, is seen the next moment to be open and changed in position.

closely to her side to steady herself. The fore-arms, but especially the right one, are in constant movement of extension, flexion, pronation and supination, and the fingers in the same manner; the contraction passing in the mode described from one to all. The muscles of the fore-arm are not hypertrophied (thus differing from Dr. Hammond's cases), but they are very firm; and though she cannot squeeze very firmly, she is yet able to resist extension of the elbow, and can feed and dress herself. Gait very awkward and tabic in form; yet the muscles of the legs are firm, though small, and she can walk about for hours without assistance. In walking she protrudes the abdomen and keeps the shoulders well back to preserve the balance, very much in the same way as pregnant women. She has good light hair in abundance, and light eyes; the ears are well-shaped and symmetrical, and the teeth regular in shape, number and position, nor is the palate high-arched, differing in all these particulars from the usual characteristics of idiots and imbeciles. The head measurements, too, are good, viz. 21 inches in circumference, 12 inches from the auditory meatus of one to that of the other across the top of the head, 13 inches from the root of the nose to the occipital tuberosity. Thyroid not enlarged, and no trace of heart disease. Sensibility of skin normal, and the intelligence fair and does not deteriorate. No history of chorea or other nervous affection of any other member of the family can be obtained.

CASE II.

H. H., female, æt. 32, of light complexion, with very good eyes; not epileptic, good teeth, ears, and palatine arch. Slight dorsal spinal curvature to the left side. Face much drawn towards the left side, and spasms of risorius, obiculares, occipito-frontalis, &c. The transverse section of the mouth is very large, and the difficulty of talking proportionately great. Her intellect is not so developed as in the last case, and though able to feed and dress herself, she cannot read or write. The waves of muscular movements in the extremities are much as those in the last case, and the tongue is protruded and withdrawn just as the head of a tortoise is pushed forward from the shell. The gait is awkward, and balance is preserved by pushing forward the abdomen. Thyroid natural; heart-sounds perfect. Head measurements good, viz. 20, 12, and 11 inches in the three directions of circumference, across, and antero-posterior. No history of chorea in the family, nor is

there any trace of delusion or hallucination; simply a congenital feebleness. The muscular movements cease during sleep.

CASE III.

L. M. D., female, æt. 17. Light complexion; can talk, feed, and dress herself, and has the use of all senses. Not epileptic. Hair very long and silky. Ears, teeth, palatine-arch, and head measurements very good, the latter being, on the above-mentioned scale, 19, 11, and 12 inches. Neither here nor in any of the other cases are there any traces of rickets. Moves her head slowly about, and the face-muscles are always on the work. No heart disease. Her sister cannot account for the condition, and says that there is no history of any nervous taint in the family.

CASE IV.

L. C., female, æt. 16. Does not speak, but can hear. Dark complexion. Teeth very good and regular. Not rickety. Head measurements are $19\frac{1}{2}$, 11, and 12 inches. Neck and head always on the move, being pushed slowly forwards and from side to side. Mouth very wide and open, owing to the retractors being always on the move, and thus a laughing expression is nearly always present; but she can cry and shed tears. Fingers and fore-arms in constant gliding motion, and the arm-muscles are very firm and well nourished. No spinal curvature. Heart-sounds good. No history of chorea.

CASE V.

M. R., female, æt. 44. Reads, but cannot write. Can feed and dress herself. Memory good, and she is generally very intelligent. Light complexion. Head moves constantly from side to side, and the fingers and toes are always going, nor can she hold her arms steadily at right angles to her body. Tongue unsteady. Head measurements very good, viz. 20·5, 12·5, and 13 inches. Legs straight, and there are no traces of rickets, but she walks with an incoherent *quasi-tabic* gait. Heart-sounds perfect, and there is no history of chorea. Sensation normal.

CASE VI.

W. B., male, æt. 24. Deaf mute; not epileptic; very light complexion. Constantly moving his head to the left side, and the left angle of the mouth and side of the face are always on

the move in the same direction. Teeth regular, but small. Good palatine arch. Very characteristic position of the hands, viz. flexion of both of them on the wrists and extension of the fingers, varied by the opposed movements. The spasm does not extend to the upper-arm. Muscles at the back of the head are much developed, owing to constant movement of the head from one side to the other. All these movements are executed with great rapidity, one after the other, when he becomes excited. The spasm extends to the toes, and he shuffles along in an awkward manner, one leg seeming to get in the way of the other. Head measurements are 21, 13, 12·5 inches.

CASE VII.

H. H., male, æt. 33. Not epileptic. Light complexioned. Is dumb, but can hear, and can use the deaf and dumb alphabet. Feeds himself, and is quite clean in his habits. Very slight, if any, curvature of the spinal column to the right side in the dorsal region. Head constantly moving about, and chiefly to the left side. Orbiculares and occipito-frontalis constantly twitching, as are also the muscles of the hands and fingers, in the manner above described. The fore-arm and the sub-occipital muscles are very firm and large; teeth good and regular, and palatine arch small. Plenty of hair on the chin and lip. Clumsy inco-ordinated walk, shuffling along, and apparently in danger of falling, though he can walk for a long time, and never actually falls. Heart-sounds perfect, and there is no history of chorea.

These seven cases clearly resemble in the peculiarity of the movements those called 'Athetosis' by Dr. Hammond; but here the likeness ends, for a summary of the symptoms in his cases (given in his own words) is: 'They come on with epileptic paroxysms, a feature accompanying other organic diseases of the brain and spinal cord. In both there are similar head-symptoms, tremulousness of the tongue, numbness in the affected side, pains in the spasmodically-affected muscles, and especially complex movements of the fingers and toes, with a tendency to distortion. There is no paralysis. The phenomena indicate the implication of the intra-cranial ganglia and the upper part of the spinal cord, and the analogies are with chorea and cerebro-spinal sclerosis. The probable seat of disease is the corpus striatum.' I prefer, then, to use the term imbecility with ataxia, leaving that of athetosis to designate those where the muscular movements come on as a disease subsequent to a

previous state of good health. The disease most closely resembling it is chorea, but the distinction is, on a careful examination, well marked. Thus in imbecility with ataxia no history of chorea can be traced, even in the other members of the family; there is no sign of heart disease, and the muscular movements differ *toto cælo* from those of chorea. In the latter there is not the wavy gliding movement resembling the peristaltic action of involuntary muscle, and the jerkings are more sudden, rapid, and unexpected, so that it is quite impossible to say which muscle will be next affected.

In ataxic imbecility one part of a muscle may be quite at rest, the other being in motion, but in chorea the whole of any muscle that is affected acts at once and suddenly. The mode of speech is another element of diagnosis; for in choreic insanity the words are jerked out in a quick; decisive manner in imbecility with ataxia they are drawled out in a manner due obviously to the slow and gradual manner in which the angles of the mouth are retracted. In both diseases the movements cease during sleep. Choreia, if long continued or existing to any great extent, leads to a condition of dementia more or less complete; but here, though there is a congenital feebleness of intellect, this feebleness does not increase; on the contrary, when educated, the subjects of it show considerable intelligence, and never descend into such a degraded form of insanity as those demented from chorea do. In its early stages the prognosis of the curability of chorea is good under the influence of well-known remedies, but in the other disorder medicines are of no use. Galvanism and tonics have no effect; indeed the general health is always excellent, and they may live a long time. A female patient in the Hampstead Asylum—a good instance of the condition in question—is more than sixty years old.

From others of the idiotic and imbecile class the line is well drawn, for in these ataxic persons the head is of ample measurements and good shape; the teeth and ears (always defective in the idiot) are excellent, and the palate is not arched, neither is there any constitutional taint of rickets, scrofula, or syphilis to be noted. The cause is at present a mystery, but of the very definite character of the group of cases there is no doubt. When an opportunity offers, the cervical portion of the cord, the nucleus of the seventh pair of nerves, and the corpora striata, should be examined, as being the lines in which any existing lesion would probably be found. The interest of these cases is that of a distinct physiological group marked by

sharp lines; for, to begin with, the facial resemblance is very strong; they might, as the appended lithographic plates show, be members of the same family; then the similarity of the movements in all the cases, the mode of speech, and so on, point to a definite central lesion of the same kind in all, whilst it is not often that we are able to isolate certain cognate specimens from a large group of congenitally-affected cases, the mind and its manifestations being so wide and various that few groupings have been possible. But, in addition, this class of ataxic persons has an interest of its own in the large amount of sympathy and patience it calls for, appearances being so very adverse. I have often been surprised at their powers of perception and memory; and the necessity of an early diagnosis is apparent, for the subjects of it are, some of them, very susceptible to their own deficiencies, which have arisen chiefly from want of education. Chorea has been termed an 'insanity of the muscles;' but if by this is meant an analogue to incoherence of speech, then the ataxia in question is again different, for the movements are by no means incoherent; they are, on the contrary, regular and constant.

It is not uncommon to meet, in asylums for the feeble-minded, with persons of fair intelligence who, owing to early epilepsy or softening, have the extremities of one side and the face muscles of the same side contracted. From the early period at which this has come on, an atrophy of the affected side has resulted, with consequent hypertrophy of the sound side. Under excitement the face is much distorted, and irregular movements, resembling the tremor of paralysis agitares, ensue. This class of cases (not a small one), when carefully examined, differs in many respects from that just discussed. They are as a rule quite up to the average of intellect of persons in their own sphere of life; and were it not for the fact that they are the children of poor parents who are unable to support and educate them, they would never be sent to an asylum. As it is, they first go to the workhouse, whence they are sometimes sent to the parish school and taught, but most often they remain in the 'imbecile wards' until an opportunity occurs for removing them to an asylum.