Journal of Neurology, Neurosurgery, and Psychiatry 1983;46:289–298

Occasional Review

The athetoid syndrome. A review of a personal series

JOHN FOLEY*

From the Cheyne Centre for Spastic Children, London, UK

"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" (Shaw, 1873).

Athetosis is a disorder of movement, not a disease However, it forms the main feature of a familiai syndrome, but its definition is difficult because our concept of the condition has expanded far beyond the terms of the original description. Some authors confine athetosis to involuntary movements of a particular kind involving the limbs. Others prefer the terms dyskinesia in relation to the limbs and dystonia in relation to the trunk,[1 2] though these terms offer no advantage in clarifying the nature of the disorder of function. Hammond, in 1871, originally used athetosis to describe involuntary movements of the limbs appearing in previously normal adults, a condition as rare to-day as "post-hemiplegic chorea". Shaw[3] described athetosis as "ataxia with imbecility" (the latter meaning in those days incapability) but emphasised its onset shortly after birth, the involvement of the facial and trunk muscles, speechlessness in some cases, and often preservation of intelligence.

The variety of synonyms makes for confusion—"double athetosis",[4 5] "Little's disease with involuntary movements", "infantile partial striatal sclerosis", "congenital chorea",[6] and "distal dystonia".[7] The greatest cause of confusion is the occurrence in some cases of signs thought to be purely "cortical". Some authors[8] feel that the term is used too loosely, that pure athetosis is rare, and that bilateral athetoid movements often complicate diplegia and quadriplegia. Many authors have referred to a few "signs of spasticity" in some cases.[3 8 9] Plum[10] was unperturbed by the occurrence of pyramidal signs in some of his cases of kernicterus, while Bobath[11] avoided a definition by stating that "most cases of the athetoid group belong to the quadriplegias".

The athetoid syndrome is here defined as a non-progressive but evolving disorder, due to damage to the basal ganglia of the full-term brain, characterised by impairment of postural reflexes, arhythmical involuntary movements, and dysarthria, with sparing of sensation, ocular movements and, often, intelligence. The layman, untroubled by neurophysiological niceties, sees the problem more simply—they can't sit, can't move at will, can't talk, and yet take everything in.

Athetoids form about a quarter of the cerebral-palsied population and, because of their latent abilities, are the cause of the emotive concern for children with brain-damage. Not only were the earlier definitions concerned mainly with involuntary movements of the limbs, but many of the reviews did not include severe cases. Some of Carpenter's[9] and Twitchell's[8] cases could walk and talk and would have been classed as mild in our present series.

Pathology

The responsible lesions are mainly in the basal ganglia[3 5 12–14] and result in most cases either from intranatal asphyxia[9] or neonatal jaundice.[10] It is curious that Carpenter[9] in his review of athetosis did not mention kernicterus, while Gerrard[15] in his study of kernicterus did not mention asphyxia as the cause of an identical syndrome. Polani[16] gave equal weight to kernicterus and abnormal labour. Plum[10] was unable to distinguish between the two varieties except in respect of deafness, though he found some "spastic" signs slightly more often in cases due to asphyxia. Exsanguination-transfusion, first done by Hart[17] in 1925, before the description of the main blood-groups, and the use of anti-D globulin, first proposed by Finn[18] and developed by Clarke *et al*[19] have now made kernicterus a rare disease, and have

Address for reprint requests: Dr John Foley, The Cheyne Centre for Spastic Children, 61 Cheyne Walk, London SW3, UK.

Received 20 April 1982 and in revised form 30 October 1982.
Accepted 20 November 1982

spared countless individuals a lifetime of misery.

The aetiological and pathological features of the diplegias and quadriplegias[20] are quite different from those of the athetoid syndrome, and the difference in pathology is reflected in outstanding differences in both the main clinical features and the commonly associated phenomena. The quadriplegias, having multiple and disseminated lesions, show in general perceptual defects due to involvement of the parietal association areas, intellectual impairment, an abnormal EEG and CT scan, and a high incidence of epilepsy. In the athetoid group, in contrast, in which the damage is almost limited to the basal ganglia, perceptual difficulties are rare, intelligence can be high, the CT scan and EEG are normal[21 22] and epilepsy is rare.

The best-known pathological feature of athetosis is marbling, or état marbré, of the basal ganglia in cases due to abnormal birth (presumably with asphyxia). Originally described by the Vogts[12] as a congenital malformation, it consists of a coarse network, or even bundles, of myelinated nerve-fibres disturbing the normal radial pattern of the striatum. Denny-Brown[6] denied the relationship to asphyxia postulated by the Vogts, while Alexander,[14] concentrating on the abnormal myelinated fibres, denied the occurrence of significant cell-loss or gliosis, though he pointed out the sharp watershed running through the striatum between the vascular territories of the anterior choroidal and the striate branches of the middle cerebral artery.[13] Alexander was emphatic that the marbling fibres ("tractus marmoratus") come from the frontopontine tract and are of prenatal origin, and are thus not ascribable to intranatal factors; Byers[23] refuted him. Cell-loss, if not gliosis, is widespread, and the globus pallidus may even show "status dysmyelinatus". An alternative is "état fibreux", a diffuse loss of nerve cells giving a false impression of hypermyelination. Marbling of the striatum is sometimes associated with abnormalities elsewhere in the brain, ulegyria or periventricular softening. Though marbling may occur near vascular lesions in adult brains, widespread marbling of the basal ganglia has no counterpart in the pathology of later life. Generally speaking the striatum is marbled while the thalamus is spared. An inherited factor may be involved as there is occasionally a familial incidence. Other aetiological factors are responsible in a proportion of cases. Malamud[24] described cases following meningitis in infancy, and 2% of our cases had this history. Of Christensen and Melchior's[20] six cases of the athetoid syndrome, including three with rigidity, all had a history of perinatal difficulties, and four had been jaundiced, but one, with a similarly affected sister, had a major central malformation.

Though the asphyxial and kernicteric varieties seem clinically indistinguishable, the distribution of lesions is different. In the former, the caudate nucleus and putamen are most involved, and this has also usually been the case in the athetoid syndrome following rotational head-injuries in childhood.[25–27]

In kernicterus, due usually to Rh-isoimmunisation, and less commonly to ABO incompatibility or glucose-6-phosphate-dehydrogenase deficiency, there is characteristic yellow staining of the dead nerve cells followed by gliosis and poverty of myelination. Schmorl,[28] who coined the term, considered involvement of the caudate nucleus to be rare. The putamen is less severely damaged than in état marbré, and the cortex is rarely affected. Schmorl found the main lesions in the globus pallidus, subthalamic nucleus, cornu ammonis, dentate nucleus, inferior olives, vermis, flocculus, and the cranial nerve nuclei of the fourth ventricle floor; to which Gerrard[15] added the putamen and caudate. Gerrard found the vestibular nuclei as often affected as the cochlear. The crucial level of unconjugated bilirubin necessary for the causation of kernicterus is 18 mgs.% or 308 μmol/l, though a lower level is injurious if the infant is premature, hypoxic, anaemic or febrile. Gerrard's aphorisms are noteworthy: "no kernicteric infant is ever normal"; "it is to those who feed, handle and hold these infants during the first ten days of life, rather than to those who subject them to a full neurological examination that, in mild cases, the presence of kernicterus is revealed." He was aware of high-tone deafness as an important accompaniment of kernicterus but was wrong in supposing that affected children are usually mentally subnormal. One of his mothers commented "the deafness is so perceptive you can't perceive it".

Athetosis also occurs in many other conditions. Apart from the drug-induced dyskinesias there are a number of conditions in which the lesion is, or is thought to be, biochemical rather than anatomical, such as familial kinesigenic choroathetosis,[29–32] paroxysmal dystonic choreoathetosis,[33 34] chorea with thyrotoxicosis[35] and choreoacanthocytosis;[36] a number of metabolic disorders, usually with severe mental retardation, may also show progressive athetosis,[37] including Lesch-Nyhan disease[38] with its combination of self-mutilation and hyperuricaemia.

The pathological processes at work in the athetoid syndrome and spastic diplegia would at first sight appear to be widely different, but anatomically the relationship between them is not as obscure as might be supposed. Greenfield[39] mentioned "periventricular rarefaction" as the commonest sequel of birth injury. In a proportion of the very premature, haemorrhage occurs into the germinal subepen-

dymal matrix,[40] and its sequel, periventricular leucomalacia, involves, amongst other things, the head or body of the caudate nucleus such as would produce disorders of posture and gait, in addition to the "spastic" signs resulting from damage to the neighbouring pyramidal fibres for the legs—hence "Little's disease with involuntary movements", and the fact that many athetoids, if they do walk at all, walk like spastic diplegics.

The present series

The 165 cases reviewed here were seen by the author between 1955 and 1980 at The Cheyne Centre, and are compared with 218 randomly assembled cases of diplegia and quadriplegia. The method of history-taking was uniform throughout, and the histories were checked as far as was possible against details supplied by the referring hospitals or doctors. The age when the child was first seen ranged from 3 months to the early teens but was most commonly 4½ years. There were twice as many boys as girls (106:59); both the total numbers in each five year period and the proportion due to jaundice fell slowly in the 1970's, but dramatically after 1975, as was found in Sweden.[2] Fifty-nine (36%) were due to jaundice, and 70 (42%) to asphyxia; there were only eight (5%) in whom there was a history of both asphyxia and jaundice; chance would have resulted in a far higher figure, and it is therefore presumed that most infants with both asphyxia and jaundice die in their first year. In four cases the jaundice was due to ABO incompatibility, in three to glucose 6-phosphate dehydrogenase deficiency, and in one to the Crigler-Najjar syndrome.

The main aetiological factors in the whole group are shown in table 1 with comparable information from the cases of spastic diplegia and quadriplegia. Asphyxia and jaundice were the principal causes. Prematurity was surprisingly uncommon in the athetoids (16%) and spastic quadriplegics, but was a major factor in diplegics (53%), in whom a history of abnormal pregnancy was also more common. Five per cent of the athetoids were postmature. The incidence of twinning was about the same in the three groups, and was considerably higher than normal. The incidence of minor external malformations was lower in the athetoid group than in the spastic cases, but nevertheless well above normal though not as high as the 24% found in cases of cerebral palsy in general.[41] Of the group of athetoids in this series without a history of asphyxia or jaundice, 41% nevertheless had a history of abnormality of labour in some form; but in this group, also, the incidence of a positive family history in respect of cerebral palsy or of epilepsy was high (31%), while a third had a history of infection or dehydration after birth. Of the 59 cases with jaundice, only three (5%) had had an abnormal birth and 15 (25%) were premature. Of the cases with asphyxia, 39 (56%) had had an abnormal birth, but only eight (11%) had been premature. These figures support the contention that athetosis is due principally to asphyxia or jaundice acting on the full-term brain. These factors operating at any other time of life do not produce the same clinical picture.

The physical signs

Rigid adherence to the original definition would restrict the physical signs of athetosis to dyskinesias of the limbs and dystonia of the trunk,[1] but this nar-

Table 1 *Main causes of cerebral palsy (percentages in brackets)*

	Athetosis (n = 165)	*Diplegia (n = 100)*	*Quadriplegia (n = 118)*
Jaundice	55 (33)	9 (9)	2 (2)
Asphyxia	71 (43)	11 (11)	19 (16)
Jaundice & asphyxia	8 (5)	1 (1)	0 (0)
Abnormal birth	62 (37)	13 (13)	26 (22)
Neither jaundice nor asphyxia	39 (24)	81 (81)	97 (81)
Prematurity	26 (16)	53 (53)	21 (18)
Postmaturity	8 (5)	1 (1)	7 (6)
Abnormal pregnancy	22 (13)	30 (30)	23 (20)
Twins	8 (5)	5 (5)	5 (5)
Family history			
Cp or Ep	4 (2)	17 (17)	16 (14)
Malformations	11 (7)	9 (9)	11 (9)
Causes after birth	4 (2)	0 (0)	5 (4)
Mean birth weight	2903g, SD 733	2066g, SD 813	
Median	3047g, range 1474–4536	1846g, range 967–4000	
Mean gestational age	38·37 weeks, SD 3·53.	33·67 weeks, SD 4·67	
Median	40 weeks, range 26–44	32 weeks, range 24–42	

row definition excludes many of the principal features of the syndrome, in particular the negative features, which are as important as the positive. These negative symptoms, in the sense used by Martin[44] in relation to disease of the basal ganglia, are always present in the early stages, usually persistent, and often more disabling than the positive symptom of involuntary movements.

While the cardinal features of athetosis are the positive one of *involuntary movements of a particular kind,* together with a negative one of *postural instability,* a proportion of cases with more diffuse lesions (albeit mainly in the striatum) have an admixture of other phenomena, commonly called "spastic"; it is these that make classification difficult. Their presence was clearly admitted by Carpenter,[9] Koven and Lamm,[45] Plum[10] and Tardieu *et al.*[43]

The series was divided into three clinical groups: *"pure athetosis",* with involuntary movements and impairment of postural reactions (63 cases), *"ataxic-athetoids",* a type specifically mentioned by Tardieu *et al*[42 43] (18 cases), and *"athetoid quadriplegias"* in whom involuntary movements and impaired postural reactions were associated with some pyramidal signs (84 cases).

The series was also divisible into three groups according to severity. The term *"severe"* refers to those children who, when past their third birthday, could not sit without support, feed themselves, or move about the floor. By *"moderate"* is meant those who could sit with minimal support, feed themselves in part, and had some form of locomotion; while the *"mild"* could sit, feed themselves, and walk about after a fashion. Sixty per cent were severe, 24% were moderate, and only 16% were mild.

Table 2 lists all the phenomena encountered. Most of these will be familiar to those who handle these children in daily life.

No satisfactory explanation of involuntary movements, except those of hemiballismus[46] has ever been achieved. Several hypothetical factors were proposed by Tardieu and Tabary,[42] and the movements have also been attributed to disordered proprioception;[47] and control of reflexes by electronic feedback devices has had some success.[48] The tilting reactions are not at fault, and it seems most probable that the movements result from disturbance of those postural reactions in trunk and limbs that are the inevitable accompaniments of perturbation of equilibrium by the movement of a limb, even if this occurs within the area of the supporting base, and variously described as the counterpoising,[44] or "anticipatory postural" reactions.[49]

It is often written that the involuntary movements are superimposed on a normal motor system, implying that they disturb the willed movement. This is probably untrue, for abolition of the involuntary movements by sedation does not leave behind the capacity for normal movement. It is more realistic to say that the involuntary movements are superimposed on a motor system that has been deprived of its automatic postural stability. There is, however, an additional factor: the voluntary movements themselves are incorrectly programmed. Nevertheless, athetoids in later life have a remarkable capacity for using machines, and it appears that they can execute small ballistic movements even when a closed-loop or guided movement is impossible.

I THE NEGATIVE PHENOMENA

The negative phenomena have tended to be forgotten because in general we are taught to observe "signs"—spasticity, tremor, involuntary movements and so forth—and it is often difficult to describe a function that is absent. Head-lag and defective trunk control are mistakenly called "hypotonia" when in effect they represent the absence of the normal axis-elevating mechanism. In discussing negative symptoms and the possibility of their correction a clear distinction must be made between automatic and voluntary actions.[44] Normal head-elevation is an automatic process involving the balancing of a mass weighing 2·6–3·6 kg on a slender column, and the process is tireless after a few months' of age: the infant has won his first battle against gravity. When automatic head-elevation is deficient a voluntary

Table 2 *Principal phenomena*

I. Negative
Impairment of postural reactions (head, trunk, limbs).
Dysarthria.
Early paralysis of upward gaze.
II. Positive
1. *Release of anticipatory postural reactions:*
Involuntary movements of face, tongue, limbs, trunk.
2. *Intention tremor*
Cocontraction, "contrary movement".
Later, Dystonia and progressive rigidity.
Ultimate hypertrophy of neck muscles.
3. *Release of Brainstem and other primitve reflexes:*
Extension spasms.
Overactive Moro response.
Symmetrical and asymmetrical tonic neck reflexes ("birdwing" and other postures).
Eye-avoiding and facial-avoiding reactions.
Conflict of grasp/avoiding reactions in limbs.
Overactive bite reflex, dysphagia, "snorting".
Athetoid "dance on hot bricks".
III. Miscellaneous
Normal reflex eye movements.
Frequent high-tone hearing loss.
Normal CT Scan.
Normal EEG.
Epilepsy rare.
Perceptual problems rare.
Normal skull-chest ratio.
High-arched palate.

effort may still bring the head up, though only briefly, and visual stimuli also help. The negative phenomena—impairment of axial elevation against gravity, and of the trunk righting and rotative reactions, are an invariable feature of the athetoid syndrome; but, because of the diffuse nature of the lesion in cases of spastic diplegia, these same features may be found in most other forms of cerebral palsy. Impairment of head-elevation is rarely, if ever, due to an excess of flexor tone, as can be simply verified by feeling the neck muscles. Often these children show a phasic alternation, periods of spontaneous head-elevation lasting a few seconds being interspersed with more prolonged periods of axial collapse in which, though conscious, they are capable of nothing.

1. Postural mechanisms

In 127 of the case-notes there was reliable information about negative phenomena. Eighty-five per cent had impaired head and trunk control in their first year, 51% in the second, and 41% in subsequent years. In many cases, these negative symptoms were long-lasting and disabling. Of those cases who were reviewed after the age of 10, 62% still could not sit, stand or walk without support; those who could sit without support could also stand and walk, indicating that stability of the trunk is the prerequisite of stance and gait. In two-thirds of the cases, however, impairment of axial control and of the righting and rising reactions remained the most disabling feature of the condition: the positive phenomena, though necessary for the diagnosis, were often less important in determining the child's ability than the negative ones, and the most disabling positive phenomena were not the athetoid involuntary movements of the fingers or limbs but the release phenomena common to many cases of central brain damage.

2. Deafness, intelligence, and speechlessness

Sixty per cent of the children were severely disabled from the physical point of view, while 24% were moderately, and only 16% mildly affected. Hearing loss occurred in 81% of the jaundice cases, 9% of the asphyxia cases, and in 39% in the whole series. (Hypoxia is now recognised as a definite cause of damage to the cochlear nuclei.[50 51]) When related to the severity of the physical handicap, the highest incidence of hearing loss was in the moderately affected, and not the severely affected group.

Dunsdon[52] found that the majority of cases of athetosis had an IQ below 85, though 8% had an IQ above 115, but this was before the era of aids to non-verbal communication. Because of the difficulty of estimating intelligence even with non-verbal tests and eye-pointing, our cases have been divided into three categories—intellectually subnormal, average, and above average (IQs under 75, 76–110, and over 110). As might be expected, a low IQ tended to be associated with greatest physical severity, but there were noteworthy exceptions: 4% of our totally disabled cases, and 5% of the moderately disabled had IQs over 120. It has been said[53] that asphyxia causes more widespread damage than kernicterus and so tends to be associated with a lower IQ. The reverse was found in the present series: 31% of the asphyxia cases had an IQ of over 100, as opposed to 17% of the kernicterus cases.

Dysarthria is an integral part of the syndrome:[54] only two cases had normal speech. Speechlessness was taken as inability to produce short phrases of more than a few intelligible words in order to express needs and thoughts. Of 151 cases in which adequate information was available, 63% were in effect speechless at the age of 5. As would be expected, speechlessness tended to go with physical severity and intellectual retardation, but once again there were important exceptions, a crippling dysarthria occasionally occurring in a child whose physical disability was only slight: the converse was rare (<1%). Speechlessness at the age of 5 proved to be as common without deafness as with it. While the highest incidence of speechlessness at the age of 5 naturally occurred in the cases of low intelligence, there was a very important minority (4%) in whom speechlessness was associated with very good intelligence, and a substantial group (28%) who were speechless at 5 and yet had an intelligence that was at least normal. The highest incidence of severely retarded speech of course was in the athetoid quadriplegic group. When the analysis was restricted to cases of "pure athetosis", the proportion of speechless children with normal or high intelligence was somewhat greater. Children lacking the ability to question must inevitably be intellectually starved, and it is doubtful whether their intelligence can be accurately gauged even by the so-called non-verbal tests available, the instructions for which are necessarily verbal. From the theoretical point of view the dysarthria is important as it casts some light on the nature of the disorder. Neilson and O'Dwyer,[55] using a meticulous polygraphic technique, have shown that the dysarthria is not due to the interference of a sustained background hypertonus, the activity of primitive reflexes, imbalance between "positive and negative oral responses", or the disruption of voluntary movements by involuntary ones—all cherished theories that have held sway too long. They conclude that the dysarthria is due to faulty command at the cortical level, an explanation that can obviously be extended also to the disorder

of limb movements. Speechless though these children are at first, very few of them are truly aphasic and some achieve a high linguistic ability as is shown by the books of poetry and autobiography written by adult athetoids.

II THE POSITIVE PHENOMENA

Involuntary movements

The abnormal postures and movements were fully illustrated by Foerster[56] and have been well described by Twitchell,[8] who summarised the observations of Hammond, Shaw, and Anton.[57] It was Anton, for example, who pointed out that often an intended movement begins in the antagonist—"contrary movement". Most of the phenomena are listed in table 2. The involuntary movements are divisible into two main groups—on the one hand, continuous irregular movements of face, tongue and extremities together with recurrent postures of the trunk, and on the other hand associated or release phenomena, such as extensor spasms and the startle reaction, that occur also in patients with diffuse brain damage. The true continuous involuntary movements are slow, sinuous and irregular movements of the lips and eyebrows, or rolling movements of the tongue with or without protrusion, purposeless movements of flexion or extension of the fingers and toes, together or individually, so that they move like tentacles; and slow writhing movements of the neck and trunk. An alternation between retraction of the neck with flexion of the head, and protraction with extension, is common. These movements are augmented by anxiety, physical insecurity, and the state of being about to act or speak; they are absent in sleep (Hammond himself being the only dissenter), at complete, secure, rest,* and when attention is deeply held. Carlson[58] remarked of such patients at a film-show "they became so completely absorbed in what they saw on the screen that all involuntary movements ceased, and for the duration of the performance they seemed completely normal". Twitchell wrote that the movements can even be absent in confident assisted standing. Short of stupefaction the condition is unaffected by drugs. The cocontraction of agonist and antagonist and the wide diffusion of muscular activity have been illustrated polygraphically by Pampiglione.[59]

The second group of involuntary movements are really obligatory postures imposed by the release of the brainstem reflexes—the tonic labyrinthine, symmetrical and asymmetrical neck reflexes and the Moro response to neck extension or to sights and sounds. In the early years the athetoids and the spastic quadriplegics share many features in common. The cardinal sign of involuntary movements may be late in appearing; but swallowing is usually more disturbed in the athetoid, and the relative normality of eye movements, together with the athetoid dance and the absence of fits or microcephaly make a distinction possible. Quite often the diagnosis has to be revised as more dystonic features appear.

In addition to these release phenomena there are a number of complex synergies that are characteristic of the athetoid syndrome. Amongst these can be classed the snarl and excessive smile that belies the patient's emotions; mass movements of limbs and trunk in flexion or extension, and a number of characteristic patterns in the limbs. These include flexion-abduction-external-rotation of the thigh with flexion of the knee, dorsiflexion of the toes and supination of the foot alternating with extension-adduction-internal-rotation of the thigh with extension of the knee and foot, this alternation being seen well in the "athetoid dance"; and pronation of the wrist in association with extension of the arm and fingers, changing to flexion of the arm with supination. A common posture, and often the final deformity, is over-flexion of the wrist with slight flexion of the metacarpophalangeal joints and over-extension of the interphalangeal joints. An attempt to reach forward is often accompanied by collapse of the head and trunk. An attempt to grasp is preceded by over-extension of the fingers ("starfish-fingers")—in other words the contrary movement; often the grasp cannot be relaxed, or the hand on contact flies away from its objective and a fresh effort has to be made; or there may be alternating grasping and avoiding reactions, neither of them powerful. Grasping is often accompanied by aversion of head and eyes, the reverse of the asymmetrical tonic neck reflex posture, though occasionally the patient may use the asymmetrical tonic neck reflex pattern to facilitate flexion of the elbow once an object is in the hand. In the prone position the arms are usually trapped under the body. Crawling is impossible in even moderate cases (though all of Twitchell's cases could crawl!). In contrast to the legs, in which the placing reaction is always exaggerated, the placing reaction in the arms may be absent, the forelimbs being neatly folded, with supinated wrists, in the ventral-suspended position. In the supine position the common posture of the arms is that of abduction-external rotation-flexion in the "hands-by-ears" position, while the lower limbs are abducted, externally rotated at the hips and flexed at the knees, in contrast to the adducted-extended posture of the diplegic. The hands-by-ears posture is often initi-

*Dejerine, quoted by Tardieu *et al*,[43] stipulated that the movements must be present at rest, but no athetoid can be said to be at rest when lying on a couch. The movements are in fact absent at secure rest.

ated by a Moro reaction, and a rhythmic succession of these reactions contributes to some involuntary movements; a child in this position cannot roll or be rolled. (This positive resistance to being rolled should not of course be confused with the negative absence of the righting and rising reactions). Those athetoids who can walk tend to lean backwards, over-flex their hips, and yet make little advance because of their backward tilt; the tendency to fall backwards is seen also in many diplegics.

THE APPARENT NORMALITY OF EYE MOVEMENTS

Being speechless in their early and middle childhood, most of these children have to depend on eye movements as their only means of communication: indeed it is their eye movements that are their salvation. Without special facilities this eye-code is limited to yes-no responses and even these can be difficult for the uninitiated to decipher. Eye-pointing demands voluntary control of saccades, and one of the remarkable things about the athetoid syndrome is that eye movements are preserved when every other kind of movement is completely disorganised. In point of fact not every kind of ocular movement is unimpaired. In this series there were 106 cases in which eye movements were noted carefully, and impairment of upward gaze, including "sunset eyes" occurred in 18%; optokinetic nystagmus was impaired in 29%, and there were abnormalities of pursuit movements in 20%; in two instances the child had mistakenly been regarded as blind in his first year. Automatic saccades are probably normal in all cases. The ocular avoiding reaction is a very common and annoying phenomenon, the head and eyes being averted from the object of interest, visual assimilation presumably being accomplished by lightning-quick sampling. One of the most characteristic features of these children is the preservation of the vestibulo-ocular reflex, the visual axis remaining stable despite the head's being tossed in all directions. Squint is much less common in athetosis (12%) than in cerebral palsy generally (37%).[61]

Onset and progression

TIME OF APPEARANCE OF INVOLUNTARY MOVEMENTS

Involuntary movements can appear quite late, though our records contain only 38 cases in which the time of onset is documented precisely. In 47% they developed in the first year, in 18% in the second year, in 16% in the third year, in 10% in the fourth year, and in 8% in the fifth or subsequent years. In over half the cases, therefore, the cardinal sign of athetosis was absent in the first year, so it is small wonder that the diagnosis is often wrong.

The outstanding early features of these infants appear to be difficulty with feeding, an over-active startle reaction, opisthotonic spasms, postures dictated by the tonic neck and labyrinthine reflexes, rhythmical jaw-opening and tongue protrusion, hence serious feeding difficulties, and the "athetoid dance" when the placing reaction is elicited;[60] the athetoid dance is in fact often the earliest diagnostic sign.

The majority (85%) of our cases showed conspicuous impairment of trunk elevation in the first year, though many of these simultaneously had extension spasms. Reliable information about rigidity was available in 105 cases. Rigidity developed in the first year in 52%, in the second year in 27%, in the third year in 11% and later in 13%: rigidity therefore developed as often after the first year as during it, and in a small proportion of cases was delayed until after the sixth year. A pathological Moro response after the second year was recorded in 24% of cases, and a prominent athetoid dance in 25%, though this is almost certainly an under-estimate, the phenomenon often being taken for granted. A myoclonic flexion-jerk was a rare but definite phenomenon, quite distinct from the usual extension spasm. Quite often the extension-spasm seemed to be partly under voluntary control and to be the only movement with which, in his earliest years, the child could express himself. A rare phenomenon, but a very troublesome one, is the dystonic arm—extended, internally rotated and abducted, which makes crawling impossible; eventually these children learn to hold the offending limb or sit on it.

EVOLUTION: THE CHANGE WITH AGE

The "Little Club's" definition,[62] of cerebral palsy as a "non-progressive but not unchanging disorder of posture and movement . . . ." is particularly apt when applied to athetosis, because athetosis changes a great deal in the course of the decades.[16] Actual progression in cerebral palsy has been recognised by Hanson *et al*,[63] whose cases of acquired mild spastic diplegia or hemiplegia later developed progressive choreoathetosis, while Burke *et al*[64] and Montagna *et al*[65] have recorded the gradual progression of athetoid and dystonic signs after birth asphyxia. An analogy with some of the drug-induced dyskinesias suggests that the condition, though histologically non-progressive, may be biochemically progressive, and results from denervation-supersensitivity of receptors in the basal ganglia that have been deprived for a while after birth of their normal supply of

transmitters. Thus the negative phenomena are the most obvious ones in the first two years or so, and the positive signs appear later. In the present series floppiness preceded rigidity in 53%, rigidity appeared first in 23%, the two appeared simultaneously in 15% and floppiness was persistent in 8%. The floppiest "rag-doll" athetoids probably die in their first two years.[10] In the majority the floppy state is punctuated by opisthotonic spasm. When automatic head-elevation appears it does so in a phasic "off-off" manner suggesting the intermittent production of a transmitter.

No adult athetoid is ever hypotonic, even though many of his postural reactions remain defective. Furthermore, involuntary movements appear gradually in the course of the first few years and tend to change from rapid choreic movements to slower dystonic ones as time goes on. The neck is thin and frail in the infant and quite incapable of the mechanical task of supporting the weight of the head; in the older child and adult it is invariably hypertrophied; finally in the adult the dystonic movements combined with impairment of postural reactions result eventually in spinal deformities. Almost nothing has been written about the final deformed and rigid condition of these patients. Hiatus hernia and anaemia are common in the early years. Pain of spinal origin is common in the later, and the author has seen dysphagia, cervical myelopathy and incontinence.

Discussion

The most remarkable features of the athetoid syndrome are that while the voluntary and postural motor systems are completely disorganised there is preservation of ocular movements, sensation, and often of intelligence; similarly language is acquired though speech, in over half the cases, is impossible, and facial movements are so distorted that they can only be interpreted by familiars. There is no other condition in which an intelligent individual may be obliged to spend a lifetime deprived of the ability to communicate or move, or in which there is such a discrepancy between motor intention and accomplishment. There is no other condition in which early prediction of ability is so often belied by ultimate achievement if modern aids are provided from infancy. Those who are familiar with athetosis are in no doubt that it is imperative that in the early speechless years non-verbal means of communication[66] must be provided if, in Shaw's descriptive phrase, "imbecility through deprivation" is to be prevented.

Many of the postural reactions never develop. In the early years when automatic trunk elevation is absent, mechanical contrivances hurt; a custom-built chair is essential. In most cases a measure of axial stability is ultimately achieved though usually without protective reactions, and these patients live in perpetual peril of an undefended fall. Even the most severe cases seem eventually to be able to manage the controls of electronic aids, perhaps because of the relative preservation of ballistic movements. Walking, after a fashion, is sometimes achieved quite late in life if contractures have been prevented, and for this reason physiotherapy should never be neglected in the school years. Negative symptoms are common to all forms of brain damage in early life and are concealed by the term "delayed milestones", and it is generally the positive signs that establish the diagnosis. It is not surprising that early diagnosis is often wrong, and questionable indeed whether it is possible before the positive signs appear. The matter of early diagnosis is of considerable importance, for the development of involuntary movements could theoretically be prevented in the early stages of supposed "chemical denervation" if the diagnosis could be made with certainty in the first few months, and the missing transmitter supplied. The most useful early diagnostic clues are the history, a normal skull circumference, normal eye movements, pharyngeal noisiness, and the athetoid dance.

In the accepted sense there is no treatment. Short of stupefaction drugs are of little value, and stereotaxic surgery,[67–69] though pursued with enthusiasm, does not seem beneficial in the long term. Mitigation of the symptoms is, however, possible, though it demands teamwork. Correct handling and feeding, measures to encourage trunk stability, the careful choice and teaching of codes, and the early provision of non-verbal means of communication add immeasurably to the happiness of the patient and his family. The severest athetoids come close to the state of "de-efferented man", and the usual mistake is to underestimate their abilities.

The following are a few simple rules that have been borne in upon the author. An athetoid infant who has had jaundice must be assumed to be deaf until the contrary is proved; if his saccades are normal he should be assumed to be of normal intelligence until, after years of observation, it is proved that he is not; emotional lability is likely to be due to either postural instability or the lack of a means of communication; intelligence cannot be assessed in the speechless child until he has been provided with a non-verbal means of communication; and his ultimate physical capability cannot be forecast before he has achieved a reasonable measure of trunk stability.

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