OBSERVATIONS OF A CASE OF CONVULSIONS. (TRUNK FIT OR LOWEST LEVEL FIT?)

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THE following case is of interest from two points of view: (1) the association of "epileptic fits" with congenital syphilis, and (2) the kind of convulsion. That the convulsions occurred in a patient the subject of inherited syphilis is, of course, of great importance. It does not, however, necessarily follow that the patient's attacks were owing, directly or indirectly, to syphilitic changes in some part of her nervous system. In this paper we remark almost solely on the patient's fits.

The state of the respiratory apparatus in "epileptic" paroxysms has received little special attention, even in cases of so-called cortical (Bravais) epilepsy fixation of the chest may be overlooked. In a case of this kind of epilepsy observed by one of us there was—along with the convulsion starting in one hand and affecting the whole of the arm, with turning of the head and eyes to the same side—complete fixation of both sides of the chest; this occurred in paroxysms of the range mentioned, during which the patient was not unconscious.

One of us (J. H. J.) has suggested that, besides what are commonly called epileptic fits, there occur, in the human subject, bulbo-pontal (lowest level) fits, analogous to those experimentally produced in some lower mammals. With regard to proving or disproving this hypothesis, it is, among other things, particularly important to note where the first spasm is, or what the very first symptoms are. Are there seizures, such as would be commonly called epileptic, which

begin by fixation of the chest, or in which the chest is involved before the limbs, if these are involved in the seizures; or, speaking more generally, are there fits which begin by convulsion of respiratory muscles?

The case we are about to report is an example of fits of this sort; it is clear that, in many of this patient's attacks, the first symptoms were involvement of muscles of both sides of the neck (back and front), soon followed by fixation of both sides of the chest. The neck muscles may have been engaged by discharge of central nervous arrangements representing them as they serve respiratorily—that is to say, as they serve in difficult respiration. And, what is particularly significant, in many of this patient's attacks, those during what we shall call the First Period, the limbs were not affected at all, although the trunk muscles of both sides were strongly engaged. Were this patient's attacks bulbo-pontal fits? There is another possibility: the seizures may have been owing to a discharge lesion of some part of Horsley and Schäfer's trunk centres. There is yet another possibility. Recently Sherrington and Grünbaum have discovered, in the chimpanzee, a trunk centre in the external surface of the Rolandic cortex, between the arm area and the leg area. They write (*Brit. Med. Journal*, December 28, 1901), that they "find between the representation of shoulder and hip an area which, next to the shoulder, yields unilateral movement of the chest muscles, and, next to the hip, yields unilateral movement of the abdominal muscles." If there be a trunk centre in the Rolandic region of the human brain, the discharge lesion in our patient may have been of some elements of it. It is true that the movements Sherrington and Grünbaum produced were unilateral, but possibly an intense ("an epileptic") discharge beginning in their Rolandic trunk centre of but one half of the brain might produce bilateral convulsion of the parts they mention.

There are difficulties in the way of each of the hypotheses we have stated; we prefer, whilst giving in great detail particulars of the attacks our patient had, to refrain from speculations on the localisation of the lesion causing them. We may, however, say what the question is.

It is of little moment whether the fits are to be labelled "epileptic" or not. Admitting that the slight fits (*vide infra* 4) in which there was movement of the eyes and no fixation of the chest, offer a difficulty, the question as to the severe fits is, "Where is the lesion (discharge-lesion) which, on its occasional "intense" discharge, produces a convulsion *beginning* in muscles of the trunk (that is, in muscles of two sides of the body, the normal action of which may be and often is bilateral) and beginning in those muscles of the two sides of the body simultaneously—a convulsion in some paroxysms (first period) not involving the limbs at all, and in others (second period) involving both arms *after* the trunk, involving these limbs simultaneously; also, what is very significant, involving the muscles of the arms from above downwards, thoracico-humeral muscles to those of the hand.

We have considered it advisable to describe the fits under two headings, for two periods, because as the seizures diminished in number they became modified in character. We wish particularly to lay stress upon this division, because, although there were some slighter fits of somewhat different nature common to both periods, no attacks such as those which formed the main feature of the Second Period occurred as far as our observations went, and they were numerous in the First Period. The following order will be followed:—(1) The history of the case; (2) the results of the general examination of the patient; (3) a description of the fits occurring in the First Period; (4) a description of the slighter attacks occurring in both periods; (5) an account of the seizures in the Second Period.

(1) HISTORY.

J. K., aged 17, is the eldest of a family of nine. Her father, mother, brothers and sisters are all living and healthy; the mother had no miscarriages until the last nine months, during which time she has had two. Most of the children are said to have a "narrow bridge" to the nose and several of them had snuffles in infancy, but none of them, as far as the mother can remember, had any rash. None have had any troubles with their eyes.

The patient was born at full term and without instrumental help; she had snuffles, but no rash as an infant; she has always had a squint which is said to have been worse since the onset of the fits. She was backward in walking, not beginning to do so until 3 years old, but she began to talk at the same age as her brothers and sisters, viz., 12 months. She has suffered from whooping cough, measles and scarlet fever. During the first dentition for about two years she had "teething convulsions" at intervals, in which she is said to have been quite quiet and "looking dead"; there were never any twitchings in these attacks.

The present illness began at the age of 9 years; the fits at first occurred about once a month, gradually increasing in frequency at the onset of puberty at the age of 15 years to about one a week. After this they rapidly increased in number up to several daily. In March, 1901, she was in a condition of *status epilepticus* for ten days, followed by a period of complete freedom from fits for a month; after this time the fits again increased in number until four days before she came under our observation on December 20, 1901, when she once more passed into *status epilepticus*.

(2) GENERAL EXAMINATION.

When first seen she was having a fit about every ten minutes, with recovery of consciousness but not of much mental power in the intervals. The general examination was not completed until December 22, the time being devoted to the observation of her fits. At this time she had had no fit for half an hour and would obey simple orders and answer simple questions. She was, however, extremely fatuous, with very bad memory, and had difficulty in fixing her attention.

She is dark, with muddy, coarse skin; the forehead is large, square and overhanging, the bridge of the nose flattened and the nostrils thick and snub. The lips are coarse, thick and habitually parted, the upper teeth projecting over the lower lip. The lower part of the face is thin with pointed chin and forms a striking contrast to the large forehead. She breathes habitually through the mouth, but can blow down her nose. The tonsils are not enlarged and neither they nor the fauces show any scars of past ulceration. The palate presents a high, narrow arch. The left upper median incisor tooth shows a well marked notch and

narrows gradually from base to cutting edge (the kind of teeth described long ago by Hutchinson). The pupils are large, equal, and re-act to light and in convergence; they dilate well under homatropine and show no iritic adhesions. There is a well marked convergent strabismus due to weakness of the right external rectus muscle; Mr. Marcus Gunn agreed with us in this observation and inference. The fundi were examined under a mydriatic by Mr. Gunn after the fits had ceased. He reported as follows:—"The corneæ are hazy and there are remains of vessels running into both—undoubted evidence of old interstitial keratitis. The fundi show numerous patches of choroidal atrophy of typical syphilitic nature, and the optic discs show the usual atrophy secondary to that condition of the choroid."

The patient's *power* generally is feeble, but except for the weakness of the right external rectus muscle there is no paralysis and certainly no unilateral weakness, all *movements* are present and well co-ordinated. *Respiration* is of the type normal in her sex—that is to say her inspiration is mainly upper thoracic; there is some widening of the costal angle and slight protrusion of the abdominal wall. In the act of drawing a deep breath, that is when told to do so, the movement is mainly upper thoracic at first, the sterno-mastoids being used; but towards the end of this voluntary act the diaphragm descends and the abdominal walls are pushed forward. *Sensation* appears to be normal in every way. The knee-jerks, when the patient was first seen, were well marked but not exaggerated, equal on the two sides, but when examined two days later and at some considerable time after a fit, they were obtained only with difficulty; this was also the case after complete cessation of the fits for three weeks. There was no ankle or rectus clonus. The *plantar reflexes*, both feet, were definitely extensor in type, but after complete cessation of the *status epilepticus* they were flexor in character. The *feet* show a high arch with some extension at the metatarso-phalangeal joints and flexion of the distal joints. This was said by the mother, in answer to questions, to have always been the same.

The *lungs* showed no sign of disease; it was particularly observed that there was no emphysema and no deformity of the chest. The heart sounds were everywhere clear and without murmurs, but tended to approximate to the foetal type, that is to say, the character of the two sounds were very similar and the intervals were nearly equal. The *pulse* was regular, soft and easily compressible, its rate being about 100. Nothing abnormal was found on examination of the abdomen.

(3) FITS OCCURRING IN THE FIRST PERIOD.

The duration of each fit was very brief, and their frequency being very great particular attention was paid to different points in different seizures. We shall first give accounts of some of the individual attacks copied almost verbatim from the notes made immediately after each observation, and shall then describe an attack compounded from the notes taken of numerous fits.

Descriptions of Individual Fits.

(1) At the moment when first noticed there was tonic spasm of the muscles of the neck on both sides, back and front, of the jaws and of both sides of the face, while the thorax was fixed in a position of partial expiration. The jaws were separated so that there was an interval of about half an inch between the teeth while both masseters and both mylo-hyoid muscles could be felt to be firmly contracted, contending against one another. The eyebrows were raised and the eyes, still maintaining the convergent strabismus noted above, were directed upwards. After a few seconds the tonic was replaced by clonic spasm affecting the whole face (the angle of the mouth being drawn a little more to the left than to the right), also the muscles of mastication, the sterno-mastoids, and the muscles at the back of the neck on both sides. Here is to be mentioned a point of great importance. *No movement or spasm of any kind occurred in the arms; they remained quite flaccid and loose.* The legs were not specially observed in this fit, but there was no obvious movement. As the fit passed off several deep inspirations were taken. Immediately afterwards the patient was asked to draw in her breath voluntarily, but appeared not to understand, and merely replied, "Yes, thank you."

(2) This fit began while I (H. D. S.) was auscultating the chest, and consequently it was observed from the beginning. The stethoscope was retained over the heart and attention was also directed more particularly to the face and neck. The patient was lying at the time on her back with her head turned to the left. The first motion consisted in a slow rotation of the head to the middle position with a little drawing down of the occiput by the muscles at the back of the neck, whilst at the same time both sterno-mastoids were seen to stand out prominently. Almost at the same moment, but I think a little later, the face muscles were contracted in tonic spasm, the eyebrows raised, the angles of the

mouth were retracted a little, somewhat more to the left than to the right. The eyes, with the convergent strabismus still present, were rotated upwards, the right being mesial while the left was rotated inwards. The teeth were separated by about three-quarters of an inch, the jaws were rigidly fixed and the mylo-hyoid muscles were felt to be firmly contracted. The tongue could be seen and did not appear to be contracted in tonic spasm, and certainly was not involved in clonic spasm later.

Respiration had ceased from the time the earliest motion was observed; and as the head moved round towards the middle line, a moaning noise was produced by, as proved by holding the hand in front of the nose and mouth, air being forced out of the chest.

After a few seconds clonic spasms replaced the tonic contraction in all the affected muscles, viz., face, jaw, neck and mylo-hyoids; at this time air was sucked into the chest with a grunting noise, as if through an almost closed glottis, by each clonic jerk. The spasm in the thoracic muscles then ceased and several deep breaths in succession were taken. The clonic spasms of the face and jaw muscles continued for a few seconds after the onset of the deep breathing and ceased last in the orbiculares palpebrarum and the frontales muscles.

The heart sounds were listened to occasionally through the binaural stethoscope; at the height of the tonic stage, just before the clonic spasms began, the heart was beating at a rate which I (H. D. S.) estimated at 60 per minute. When counted shortly before the fit the pulse rate was 102 per minute. When the clonic spasms began the noise of the contracting muscles and of the indrawing of the breath prevented the heart sounds from being heard, but immediately after the fit the rate was 120 per minute. The arms and legs were not watched in this attack.

(3) The patient was lying on her right side apparently asleep. As I (H. D. S.) approached the bed there was a slow conjugate movement of the head and eyes to the left. The limbs being covered nothing could be seen of any movement or spasm, but through the clothes there did not appear to be any movement of them. The corneal reflexes were present when I (H. D. S.) reached her and the head slowly came back to the original position and the patient began to mutter inarticulate sounds. A few seconds later one of the attacks similar to those described above began. Attention was at once directed to the pulse and to the erector spinæ muscles, which could easily be observed as she was lying on her right side. At the moment of feeling the *pulse* the tension seemed higher than when examined at other times

and it distinctly increased, while the pulse rate became slower until the clonic stage was reached, when the tension markedly dropped and the rate became much more rapid.

Although the muscles at the back of the neck could be felt to be in a state of tonic contraction and later in the seizure were seen to undergo clonic spasm, I (H. D. S.) was unable to satisfy myself that there was any spasm in the muscles lower down the spinal column.

In other fits attention was directed specially to the condition of the limbs, the abdominal muscles, the degree of expansion of the thorax, the colour of the lips, and the presence or absence of sweating about the forehead.

Description of the character and course of the Fits compounded from all the observations made at different times during the First Period.

For convenience the fit may be divided into three stages: the tonic, the clonic, and the stage of recovery, although the three merge into one another and in part overlap.

STAGE 1.—Tonic spasm occurred first in the muscles of the neck, followed almost at once by spasm of the muscles of respiration, causing complete fixation of the thorax. Then followed spasm of the face, the jaw, the mylo-hyoid and the abdominal muscles; and for all these it was impossible to determine any definite order. It is to be understood that the spasm of all the regions involved was of both sides at once and, except for the muscles of the face, of both sides equally. These several muscular regions will now be considered seriatim.

Neck.—The head was rigidly fixed in the mesial position, with the occiput drawn a little downwards and backwards as a result of the contraction in the muscles in the back of the neck, while both sterno-mastoids stood out prominently.

Respiratory Muscles.—As the spasm involved these muscles there was a brief moaning expiration, evidently through an almost closed glottis, ending with complete fixation of the thorax, and thereby arrest of all respiratory movement. The degree of expansion of the chest when this arrest occurred was in most observations below the mean between full inspiration and full expiration, but on two or three occasions it was noted that the thorax was expanded rather more than the mean.

Abdominal Muscles.—These were found to contract and to become firmer and more resistant, but the degree was never great.

Face.—The eyebrows were raised and the angles of the mouth

drawn outwards; generally, if not always, more towards the left than the right side.

Jaws.—The masseters were felt to be strongly contracted, but the teeth were separated by about half an inch, indicating powerful spasm in the depressors of the lower jaw. No deviation of the mandible was noted, although on one occasion there was some doubt as to whether it was not directed a little to the left. The mylo-hyoid muscles were rigidly contracted.

Eyes.—The internal strabismus due to weakness of the right external rectus muscle was always maintained, and the eyes were strongly rotated upwards, the upper lids being also elevated.

STAGE 2.—Typical clonic spasms followed the tonic contraction in all the affected muscles, including in some attacks, but not in all, the respiratory muscles; when they were so affected air was sucked into the chest by each spasm, causing a grunting noise. The abdominal muscles were never observed to undergo clonic spasm.

STAGE 3.—*Recovery.*—As the fit passed off, the spasmodic fixation of the thorax ceased first and there then ensued one or more deep breaths; it was considered probable that the depth of inspiration was greater at this time than when the patient was asked to draw a deep breath during an interval between two attacks. On several occasions she was told to draw a deep breath immediately after a fit was over, with a view to determining whether or not there was any weakness of the respiratory muscles as a sequence of the previous spasm—such weakness as is seen in a limb after a seizure due to (remaining after) some local cortical discharge. But on no occasion could she be induced to draw in her breath when told; the shortest interval between a fit and the performance of this voluntary act was about five minutes. The non-compliance with this request was probably due to the mental state, as she made no attempt to make the movement and generally answered "Yes," or "Yes, thank you." With regard to the order in which cessation of spasm occurred in the other muscles, the only observation on this point is recorded above when the last muscles to relax completely were the frontales and orbiculares palpebrarum.

The *pulse* was observed to become slower in rate but of higher tension up to the time when the tonic spasm of the thoracic muscles gave away; there was then a notable and rapid fall in the tension and a marked increase in the frequency leading to a more rapid rate than had been present before the seizure.

Reflexes.—The corneal reflex was definitely lost and probably

unconsciousness was complete. The knee jerks became a little more marked during a fit, but were never much exaggerated. The plantar stimulation yielded extensor responses which continued to obtain between the attacks until the *status epilepticus* ceased.

So far we have dealt entirely with the positive results of the observations, but there still remain the equally important negative results. The *limbs* were very carefully examined, the sister examining the thighs both with hand and eye in many attacks, and we are confident that *there was no spasm of muscles and no movement of the limbs of any kind whatever at any time during the attacks*; both arms and both legs remained absolutely flaccid throughout the seizures. *No definite change of colour* in the face or lips could be determined, although frequently looked for, and there was *never any sweating* of the head or face either to be seen or felt.

The duration of the fits varied a little, but may be stated roughly to have been forty to sixty seconds. When the fit was over the patient remained in a heavy, stupid condition for some minutes and often until the onset of the next attack. She would, however, sometimes obey simple commands, but never spoke rationally, her almost invariable reply to any remark being "Yes" or "Yes, thank you."

(4) SLIGHTER FITS OCCURRING IN BOTH PERIODS.

On the fourth day after we first saw the patient some very short fits were observed for the first time; they continued to occur with considerable frequency from that time onwards. One of these is briefly described above as occurring just before the onset of fit 3 in our account of the individual fits of the First Period. These slighter seizures were very brief, lasting only a few seconds and consisted in a slow conjugate deviation of the head and eyes; this movement was generally towards the left side, but on several occasions it was towards the right and then the convergent strabismus was still maintained, the right eye moving only a little beyond the median position, whilst the left well into the inner canthus. A very fine symmetrical rotatory nystagmus was seen in the eyes in some of these slight attacks; and flushing of the conjunctival vessels with some lachrymation was also noted. There was no fixation of the thorax in these attacks, although on one or two occasions it was thought that respiration became a little slower, but of this we are not certain. No spasm was observed in any other muscles.

(5) FITS OF THE SECOND PERIOD.

The first of this series of attacks was seen on the sixth day after the patient came under our observation, and even at the risk of being accused of too much repetition we must reiterate that no attack such as that described below, that is with convulsion extending to the limbs, was observed during the First Period. The seizures in the Second Period differed from one another only in severity, hence it will be unnecessary to do more than give one description compounded from all our observations, which were not nearly as numerous as in the case of the fits in the First Period because the frequency became progressively less.

The first change observed in these attacks was a sudden and complete fixation of the thorax with arrest of all respiratory movement; this was followed by spasm, on both sides of the body, of the muscles of the face, jaw, neck and upper extremities, and in some cases, but not in all, also of the muscles of the lower extremities. The spasm, at first tonic, very shortly became clonic. The head was retracted and there was sometimes conjugate turning of the head and eyes to the left, although both sterno-mastoids contracted. The upper extremities were not affected until the tonic contraction in the muscles of the face, jaw and neck had given place to clonic spasm; *the order in which the muscles of the arms were involved was from the large thoraco-humeral muscles to the smaller forearm and hand muscles.* The trapezii seemed to be the first to contract, and this was followed almost at once by contraction of the deltoid and the remaining muscles of the shoulder girdle. Next followed spasm of the upper arm muscles, and finally those of the forearm and hand were involved. The spasm was at first tonic and resulted in adduction with slight flexion of the shoulder and slight flexion at the elbow and wrist with pronation of the forearm. The fingers were flexed at all joints and closed over the thumb, which was bent into the palm of the hand. In some attacks the left arm appeared to suffer more than the right. This tonic stage was rapidly replaced by clonic spasm of small range and brief duration. The lower extremities were unfortunately never very carefully observed, but the movement in them was never great. Spasm was felt in the thighs and in some attacks slight flexion of the hip and knee occurred. The position of the feet we are unable to describe owing to imperfect observation.