Cæsarean Section in a Case of Myasthenia Gravis, Gravida.

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Myasthenia gravis is a disease of which an increasing number of cases is being recorded, and physicians have now tabulated a sufficient number to establish the disease as a clinical entity.

This disease attacks both sexes, and the question for us to consider, when it attacks women, is, particularly in reference to pregnancy and labour, or their treatment if that physiological condition should supervene upon this admittedly grave disease.

Briefly the clinical feature of the disease is weakness of some or all of the *voluntary* muscles, sometimes amounting to paralysis; after a prolonged rest, these same muscles may respond to the will, but again rapidly become exhausted.

The affected muscles often exhibit the "myasthenic reaction," becoming exhausted by faradic stimulation, *i.e.*, stimulation of the nerve endings, just as they are by voluntary effort.

Facial paresis and ophthalmoplegia give a characteristic appearance; and there is observed a *variability* of the symptoms, and an absence of muscular atrophy and of the reaction of degeneration.

The muscles most constantly in use are those most commonly implicated, and the bulbar muscles are very generally involved.

Males and females are alike attacked, and the symptoms follow upon some previous affection; influenza, exanthemata, emotional excitement, over-exertion, etc.; and in *women*, menstruation and pregnancy.

There are no sensory symptoms, and the prognosis is grave, death occurring in a large proportion of the cases, but without any pathological, structural changes.

Hun published an extract of 114 cases, with 50 deaths. Attacks of dyspnœa, and death from respiratory failure, make the prognosis so grave.

Dr. Warrington, to whom I am indebted for this patient, published the following description of the case in the *Medical Chronicle* of August, 1904:—

"C.M., a married woman, æt. 25, came to the hospital on the 25th February, 1904, complaining of progressive weakness in the limbs

which had first become noticeable about 18 months previously, the patient then being pregnant at an early period. The weakness rapidly progressed, and was very marked when the child was born. For some months the patient had noticed a thickness and difficulty in her speech; there had also been trouble in swallowing with occasional regurgitation of fluid through the nose. She was a well-nourished woman, with a healthy complexion. The attention was at once arrested by the peculiar aspect of the face, characterised by a fixity of expression; the lips were constantly kept just apart, the neck bent slightly back and the upper eyelids somewhat drooping. In speaking it was obvious that a rather distressing effort was needed, the words at first being indistinct and nasal; it seemed as if there was an initial difficulty in closing the posterior nares by the soft palate. The attempt to swallow was also attended by painful spasmodic movement. The aspect at once suggested the condition of myasthenia, and I will now give a description of the symptoms:—

"The gait is slow and rather waddling in character, recalling the gait of pseudo-hypertrophic paralysis. The patient very soon tires, she can only walk about ten yards, and requires a little support to prevent herself from falling. She is unable to raise herself from a chair to the standing posture when the arms are folded across the chest, and one knee cannot be lifted over the other; the movements of the leg and foot are, however, fairly strong, the weakness being chiefly in the ileo-psoas and gluteal groups of muscles. The muscles are well developed without any wasting. The knee-jerks are brisk and do not tire; they were elicited one hundred consecutive times without any enfeeblement in the response. The ankle-jerks are present, and the plantar reflex is flexor in type. In the upper limbs the weakness is even greater; the grasp is very feeble and after several attempts disappears altogether, the fingers hardly closing on the observer's hand. Abduction of the arm to the right angle cannot be performed, though it can be raised for a short time in the sagittal plane and held out against feeble resistance; elevation to the vertical line is impossible. The other movements of the shoulder girdle and forearm are also feeble. The deep reflexes are normal. The muscles of both limbs react naturally to electricity, and do not show the 'myasthenic reaction' in which faradism soon fails to cause contraction whilst the constant current remains capable of stimulating the muscle fibres. The trunk muscles are also weak, but the inter-costals and diaphragm act well, and there is also good power in extension and flexion of the head. It is, however, in the face and eyes that the most striking anomalies appear. Owing to weakness in the orbicularis oris the lips

cannot be closed though they can be slightly approximated towards each other; paresis of the buccinator prevents any distension of the cheeks, and in smiling there is elevation alone of the angle of the mouth. If in attempting to smile a knitting needle is held vertically at the angle of the mouth, it is seen that there is no outward movement at all. The risorius and zygomaticus are paralysed. Sir William Gowers describes the movement as a 'nasal snarl,' a graphic description for this striking appearance. With the paresis of the orbicularis oris there is an inability to contract the transverse muscles of the tongue, the two movements always being associated in action and probably innervated from the same cranial nucleus. There is ptosis, and the action of the levator palpebræ rapidly tires, so that whilst at first the pupils are visible, after a few attempts to raise the lids the effort becomes ineffectual, and the lids gradually drop until the pupils are covered. With repeated attempts an endeavour is made to compensate this weakness of the levator by greater extension of the head, whilst the frontales after a few feeble contractions remain motionless. When the patient is asked to gently close the eyes a narrow slit of sclerotic is still seen, and there is very feeble power of keeping the eyes shut against resistance. The pupils are equal and react to light and accommodation, so that the patient can read Jager 1 at 12 inches. The eyeballs converge equally. The associated lateral conjugate movements are deficient in both directions, the upward movements less so. The limited range of these movements can be shown by a diagram similar to that used by Sir William Gowers. It is further to be noted that such limited upward movement as there is in the left eye, is always associated with some deviation towards the inner side, and that a similar deviation inwards occurs on looking downwards. This would seem to indicate a special weakness in the superior and inferior oblique eye muscles. Fatigue is readily produced in the lateral conjugate movements, so that after repeated effort the range of movement is reduced to nearly one-half the original excursion. The muscles of mastication are distinctly weak, and become more so after effort. The palate moves rather sluggishly, though the range of its excursion does not seem to be lessened by repeated effort. The patient can repeat the physiological alphabet without any recognisable defect, but in talking it is often noticed that there is the initial difficulty already alluded to. The vocal cords move equally, but the impression is gained that the range of adduction and abduction is smaller than is natural. The electrical reactions of the facial muscles are important. As compared with a normal person, they are much less

excitable whether a large nerve branch is stimulated or the 'motor points,' in particular it is impossible to succeed in drawing the angle of the mouth outwards. The muscles involved in this movement therefore give a partial reaction of degeneration. The orbicularis palpebrarum and orbicularis oris contract fairly and more movement is attained by this means than the patient is capable of producing by voluntary effort. The other facial muscles react fairly and the myasthenic reaction cannot be seen.

"The patient has now been under observation six months, and has been kept chiefly at rest in bed and given hypodermic injections and strychnine. There is undoubtedly some increase of strength, but in all essentials the condition has remained stationary, and though some movements can be performed which on admission were impossible, fatigue quickly follows, and the myasthenic reaction has appeared in the deltoid and biceps."

Soon after admission to the Northern Hospital, it was found that she was pregnant; and as pregnancy advanced the disease became worse, until it was impossible for her to perform the most trivial acts for herself, without causing the greatest exhaustion, and the onset of alarming dyspnœa.

Dr. Warrington kindly sent her to the Lying-in Hospital, on October 12th, 1904, the confinement being expected about October 20th. On admission, her condition was one of pitiful weakness and exhaustion. In the first 24 hours she had four attacks of dyspnœa, each lasting five minutes, and for 48 hours there was no sleep.

Dr. Warrington pointed out the extreme gravity of her condition, and that these attacks of dyspnœa might end in sudden death. He feared the effect of the advent of labour, and was satisfied, that if she did not succumb in the first stage of labour, an attempt at the use of the accessory powers would almost certainly bring on a fatal attack of dyspnœa; that chloroform would most likely have to be administered, and that the use of that drug was also attended with the greater risk of respiratory paralysis.

Parturition, being a physiological act, which even when associated with the most serious systemic troubles, rarely seems to aggravate them, I was of the opinion that the anxiety as regards the *effect* of parturition, was somewhat exaggerated, and determined to wait for the natural commencement of labour and act accordingly.

On October 17th, after having slept only 2½ hours in 48 hours, dyspnœa came on at intervals and the respiratory movements were

confined to the sterno-mastoid, sterno-hyoid, sterno-thyroid muscles, and the diaphragm; pulse 90 well filled and regular, cyanosis marked.

October 18th, in spite of a good night's rest induced by paraldehyde the dyspnœa was worse, and the only respiratory movements were hiccough.

I now realised the extreme gravity of her condition, and the possibility of death from respiratory paralysis, undelivered or during delivery.

Emotion, having a marked effect, and the patient having been led to dread the advent of parturition; the risk of chloroform, either with natural labour or manipulative interference, led me to decide upon Cæsarean section, as being the procedure which we could best rely upon for a speedy termination of the pregnancy, in a manner which could be most under our own control.

The husband was informed, but not the patient, and on October 19th, with all preparations ready and all waiting, she was brought in, anæsthetised, and the operation completed without difficulties in 45 minutes from commencement of anæsthesia. Recovery was uninterrupted and gradual improvement as regards the respiration soon showed itself. During the puerperium gradual improvement has taken place in the myasthenia, but she is still helpless. The child is a well-nourished, healthy female.

I have been unable to find any reference to the effect of parturition in advanced cases such as this; pregnancy is mentioned as an exciting cause, and in this case the disease commenced during a former pregnancy. One patient with ocular paralysis became pregnant, and got practically well of the paralysis; one woman developed the disease when six months pregnant, improved and then three years later is recorded as a typical case of myasthenia. Two cases commenced after confinement.

Thus the effect of pregnancy does not appear to be constant; generally it is an exciting cause; on the other hand, there is evidence of its having been curative, but the balance of evidence seems to be that pregnancy increases the disorder.

The distress of this woman for weeks was so very great and the imminence of fatal symptoms so alarming that I should advocate the emptying of the uterus in such cases in early pregnancy, and if, as in this case, the pregnancy was allowed to go to term, considering the *variability* of the symptoms, would await the onset of labour and be guided by the condition of the patient, unless serious symptoms supervened.

After leaving the Lying-in Hospital she was under the care of

another Liverpool physician for some weeks, who reported very slight improvement in the disease with the exception of freedom from attacks of dyspnœa.

Pathological changes have not been found in this disease; it was thought that there might be some change in the muscular tissue; sections of the uterine muscle and of the rectus abdominis muscles were taken at the time of operation, but the microscopic examination of those tissues Dr. Warrington reports as negative.