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Original Papers.

THE PROGNOSIS AND THE LATE RESULTS OF POSTENCEPHALITIC RESPIRATORY DISORDERS.*

BY W. ALDREN TURNER AND MACDONALD CRITCHLEY, LONDON.

INTRODUCTION.

THE disorders of respiration associated with epidemic encephalitis have been the occasion for a considerable literature, mostly of a descriptive nature. Efforts have also been made to indicate a pathogenesis, although the problem cannot be said to have emerged from the stage of hypothesis. There has hitherto been no effort to trace the end-results of patients suffering from these respiratory disorders, and consequently no certain prognosis has been applicable.

In a previous work on the respiratory disorders in encephalitis Turner and Critchley¹⁴ modified the original classification of Marie and Lévy¹², and referred to (1) disorders of respiratory rate, (2) disorders of respiratory rhythm, and (3) respiratory ties. The first group included cases of tachypnoea and of bradypnoea; in the second category the cases of breath-holding spells, apnoeic pauses, and Cheyne-Stokes respiration were placed. The third group comprised such disorders as spasmodic cough, involuntary yawning, blowing, sniffing, etc. Dealing with the problem of prognosis, the authors state: "two main questions naturally arise . . . first, is the appearance of respiratory symptoms of itself a feature of serious import? And, secondly, how long do the respiratory disorders persist? . . . There is a certain amount of evidence to suggest that the appearance of tachypnoea . . . is of grave outlook in the older patients, particularly when associated with pyrexia . . . The duration of these symptoms seems in the majority of cases to be indefinite, although now and again a case will improve spontaneously. . . . The majority of cases however tend to persist with unabated violence for an indefinite length of time. They show no tendency to progress, however; this prognosis apparently holds good for all the varieties of respiratory disorders."

Such was the position in the spring of 1925: since that date other cases have come under observation, and many more isolated case-reports have been published. Some of these latter have been recorded as "cured" by some

* Work carried out under the auspices of the Medical Research Council.

particular therapeutic measure, such as freezing of the phrenics (Gamble *et al*⁵), by psychoanalysis (Witzel¹⁶, Jelliffe^{8,9,10}) or by hypnosis (Runge¹³). The object of this paper is purely utilitarian: its aim has been to determine if possible the ultimate fate of patients showing this particular variety of sequela. The cases originally reported by us have been followed up and to these have been added others which have come under our personal observation or been reported to us through the kindness of other physicians.

We have not included in this paper acute cases in which respiratory manifestations were present. Hall's⁶ conclusions with regards to these patients will be referred to.

LITERATURE.

The literature of encephalitis, despite its magnitude, contains surprisingly little reference to the prognosis of the respiratory syndromes. A few isolated reports can however be traced in which cases of this variety of sequela have been followed up over some length of time.

The first instance of postencephalitic respiration anomaly quoted in the United States was reported by Aronson¹ (1921). His patient, a boy of 8, showed tachypnoea and dysrhythmia: the author propounds the query as to the ultimate fate of this child: although the case is quoted by Turner and Critchley as having cleared up in six months, such is apparently not the fact, for in a personal communication to Jelliffe made in 1926, Aronson stated that there was no recovery, and that the boy had developed "some interesting regressive character anomalies."

In 1922 Mlle. Lévy¹¹ reported in her monograph *Les manifestations tardives de l'encéphalite* numerous examples of respiration disorder. Case 35 of her series was that of a woman of 35 who developed tachypnoea and Parkinsonism after acute encephalitis in 1920. After 30 months there was an apparently perfect recovery. Another patient (Case 37), a male of 29, developed a "puffing" respiratory tic, which was still present two years later. Case 62 concerned a boy of 10 who developed blowing and sniffing tics which came on in paroxysms. Each attack brought about such exhaustion that he dropped off to sleep. His conduct deteriorated so much that he could no longer be retained at the Salpêtrière but was transferred to St. Anne and later to the Vanclure. His respiratory tic was still present two years later. Her last patient (Case 67), a girl of 14, was admitted to the Salpêtrière nine months after her acute attack, with Parkinsonism and a tic-like cough. Polyuria and polydipsia were also present. Her respiratory tic still continued at the time of publication, viz., nine months afterwards.

Runge¹³ in 1923 gave a very full report of a case of postencephalitic Parkinsonism with tachypnoea and tetany arising in a young student. The case was investigated from a psychological standpoint and inherent psychoneurotic trends were revealed. Psychoanalysis exposed a masturbatory compulsion and under hypnotic treatment over a period of one year the patient definitely improved.

In the same year, Fiamberti⁴ reported the case of a girl of 12 who after encephalitis developed Parkinsonism and paroxysmal tachypnoea. At the onset of each attack the child would bend forwards nearly double and then abruptly rise erect, clutch her throat and give a forced expiration. The sphincters frequently became relaxed. Attack would follow attack until she ultimately fell into a sleep of exhaustion. Eleven months after coming under observation she fell ill with paratyphoid fever; during her convalescence the paroxysms of disordered breathing abruptly left her and did not return.

At the 1923 Conference of the British Medical Association Hinds Howell⁷ mentioned the case of a female patient, age 20, who developed dyspnoea after encephalitis. This symptom after persisting for three weeks improved but later relapsed. At the time of announcement the respiration had become normal, though the length of time after the original illness was not stated.

Wimmer's monograph on Chronic Encephalitis¹⁵ (1924) contains an account of a girl, age 18, who showed attacks of yawning. Gradually these spells left her, only to be replaced by paroxysms of tachypnoea, severer in nature than the original disability.

Witzel¹⁶ (1925) records the histories of two patients: the first one, a girl of 14, developed character changes and rapid laboured breathing after encephalitis. Under psychotherapy the respiration became normal. His other patient (Case 4), a boy of 13, was subject to spasms of noisy panting respirations. There was no history of acute encephalitis, and no mention is made of the presence of any abnormal physical signs. This patient also became well after psychotherapy.

In the same year, Gamble, Pepper and Miller⁵ gave details of a patient of 18, who developed a severe tachypnoea one year after an attack of encephalitis. There were other evidences of central nervous disease, including a right-sided hemianæsthesia, weakness of the face, nystagmus, and sluggish pupillary reactions to light. With a view to checking the tachypnoea, both phrenic nerves were exposed in the neck and blocked by freezing with ethyl chloride. During this operation the respiration rate fell from 96 to 20 per minute. No further tachypnoea occurred up to the time of publication in October, 1925 (*i.e.*, nearly seven months).

A case of paroxysmal breath-holding reported in 1925 by Hall, Mayers and Kerr¹⁷, concerned a youth of 19 who developed encephalitis in 1920. He first came under treatment for his respiratory disorder in 1923; there was no Parkinsonism. He left hospital on July 8th, 1923, but returned the following February with a well-marked adiposogenital syndrome. During this interval the attacks of breath-holding had entirely ceased. The respiration remained normal up to the time of publication.

One of the latest additions to the literature is contained in the Medical Research Council's report on the 1924 epidemic of encephalitis in Sheffield (issued in 1926). In their report Hall and Gurney Yates⁶ investigated the prognostic significance of various of the sequelæ: they dealt *inter alia* with

respiratory manifestations and considered them separately, according as they arose during the acute phase or as an after-effect. Of the former group of respiratory disorders they stated: "There is a distinct suggestion that its presence in the primary attack is of somewhat serious significance as regards life. Thus only 33 per cent. have recovered, 28 per cent. have not recovered and 39 per cent. died. . . . This mortality is more than double that in the cases as a whole, and exceeds considerably that of any other single symptom amongst those investigated. . . . Of the 18 cases in which the breathing was disturbed in the acute attack, not one showed any similar or other respiratory abnormality at the end of 1925." Regarding the second group of cases (numbering 20 in their series) they state: "Late or residual disorders of the respiratory mechanism are not usually preceded by anything similar in the primary attack. They are not of such serious importance as regards life." The fate of the patients *qua* respiratory disorder is not dealt with.

Jelliffe^{8,9,10} reported on several occasions an account of a postencephalitic respiratory disorder occurring in a male of 21; polydipsia, polyuria, character changes and Parkinsonism were present in addition to spells of tachypnoea and breath-holding. In some of these attacks, tetany would develop. Under psychoanalysis—carried out over a period of six to seven months—the paroxysms ceased and remained absent for a period of 12 to 18 months (*i.e.*, up to time of publication in January, 1927). We were informed by Jelliffe in October, 1927, that the breathing has remained normal and that some improvement is noticeable in the degree of Parkinsonism.

In the Annual Report (1926-1927) of the Metropolitan Asylums Board Borthwick² publishes interesting details of some cases of respiratory disorder treated in the Encephalitis Unit of the Northern Hospital, Winchmore Hill. Amongst these is the case of a girl of 14 who suffered from frequent paroxysms of breath-holding; during the period of inspiratory apnoea the diaphragm descended to an extremely low level, and the heart became grossly distorted. There were no physical signs of Parkinsonism. The child—whose acute attack had occurred in July, 1924—had been a patient since February, 1926; when observed in October, 1927, *i.e.*, 20 months after her admission, the respiratory attacks were still present with unabated violence and frequency.

PERSONAL CASES.

The material upon which the present analysis is based comprises primarily the original seven cases reported in our first paper. Unfortunately one case in this series (No. V) has been lost sight of. In addition we have dealt with 23 other cases, most of which have occurred in the practice of the National Hospital, Queen Square. For several of the cases we are indebted to the courtesy of Professor Hall, of Sheffield; through the kindness of Dr. Feiling, Dr. C. P. Symonds, Dr. Pearson and Dr. Greig Anderson we are also able to include those of some other patients.

We have considered the outcome of these cases under five main heads, viz. :—

- A. Cases of complete cure.
- B. Cases in which respiration has become normal although the mental or psychical concomitants are unchanged or worse.
- C. Cases in which respiration has improved, although there has been no corresponding alteration in the associated manifestations.
- D. Cases in which respiratory disorders persist unchanged.
- E. Fatal cases in which death occurs during the phase of respiratory disorder.

A. *Cases of apparent cure.*

We have only met with two cases in our experience which can be regarded as a cure, in that all respiratory symptoms have disappeared, and there have been no mental or physical sequelæ characteristic of encephalitis in their place. Our first case was actually one of the severest cases of paroxysmal breath-holding which we have ever encountered, and was reported in full in our original communication.

1. H. B., male, age 16 (patient of Dr. Turner, at Queen Square), had a mild attack of acute encephalitis in April, 1924. He came into hospital in the following September because of frequent attacks of breath-holding, accompanied by cyanosis, mental obfuscation, and many bizarre grimaces and contortions. There were no character changes and no physical signs of Parkinsonism. He remained in hospital six months, during which time no marked change occurred either in the frequency or character of the respiratory disorder. A week after discharge he was sent to the Berkshire Mental Hospital, having developed auditory hallucinations. He believed he heard the devil whispering to him and telling him to hold his breath.

He was seen by one of us at the asylum in November, 1925. He had lost a good deal of weight; his breathing was noisy, rapid and laboured, but the breath-holding paroxysms were less severe and less frequent. He complained of pain in the right shoulder and inability to raise the arm above a right angle. The right hand was wasted, but there were no objective sensory changes.

Christmas, 1926, while in the asylum, the patient developed a severe attack of influenza. On recovery his breathing was almost normal and he since has gradually recovered. He rapidly regained weight, and his delusions and hallucinations left him entirely.

He was seen again by one of us in May 1927, when his breathing was perfectly normal: he looked well, and discussed his previous mental illness candidly and freely. He was still unable, however, to raise the right arm above a right angle. There was no trace of Parkinsonism.

In August 1927 (*i.e.*, exactly three years after the onset of respiratory symptoms), he was again seen. He was still weak in the right arm; there was no wasting, but there was an area of analgesia embracing the whole of the right upper arm, the right shoulder, and the right side of neck, and thorax, back and front. There was a sharp line of demarcation at the edge of the chin and at the elbow joint. The condition was obviously a hysterical one and under gas the shoulder joint could be freely manipulated in all directions.

The completeness of the recovery in this instance was almost dramatic. His paroxysms during the active stage were of such severity and frequency as to suggest an outlook of the gravest possible type. He was becoming progressively weaker and rapidly losing weight. The disappearance of respiratory

symptoms during a severe febrile illness recalls the similar case of Fiamberti, in which the breathing became normal after an attack of paratyphoid fever. A feature of extreme interest lay in the appearance of a paralysis of the arm ; although probably arising in the first place as the result of trauma, such as might be occasioned by the excessive contraction of the accessory muscles of respiration, the condition of the arm when last seen was clearly one of hysteria. There was no limitation of passive movements under anæsthesia, and in addition there was a wide area of analgesia which was sharply delineated and corresponded with no known neural segmentation. The patient has no retrograde amnesia ; he can discuss without emotion the details of his hallucinosis.

2. H. L. H., male, age 16, came under the care of Professor Hall in November 1924. His acute attack had occurred in the previous February. When seen in November he had no evidences of objective change in his nervous system, but complained of frequently recurring "blowing" attacks, accompanied by nausea and vertigo. In the severer attacks tetany supervened, and though there was no actual loss of consciousness some degree of mental confusion occurred.

His physical state in February 1927 (two years after the onset) had not changed, but the dysrhythmia had ceased. For the past year he has been in continuous daily employment, and has been attending in addition a night school three evenings weekly.

B. *Cases in which respiratory disorder has ceased but other physical or mental sequelæ have appeared or become worse.*

3. V. W., female, age 15, was admitted to Queen Square under the care of Dr. Birley in November 1924. There was a history of an acute attack of encephalitis in the previous April. Whilst in hospital she exhibited an inverted sleep rhythm, slight twitching of the corner of the mouth, and involuntary protrusion of the tongue. There was spasmodic torticollis and a very mild degree of left-sided Parkinsonism. At night time her respiration became noisy, rapid and irregular, being interrupted by periods of apnoea.

She left hospital in January 1925, but her mother could not keep her at home because of a gradually increasing change in disposition. She "turned against her little sister," threw a chair at her mother, and threatened suicide.

In January 1926, she was admitted to West Park Asylum, Epsom. She was there visited by one of us at the end of July 1927 (two years and eight months after first coming under observation). She then showed a moderately severe grade of Parkinsonism, especially on the left side. There were no longer any involuntary movements of the face, tongue or neck. Mentally she was childish, and very quarrelsome. She was retarded and becoming progressively slower in cerebration. She exhibited an abnormal craving for attention and sympathy, even to the length of inflicting bruises and cuts upon herself.

No respiratory disorder whatever was complained of by her or noted by the attendants, either at night time or by day.

4. E.S., male, age 19 (case of Dr. Collier, at Queen Square), first came under treatment in January 1925. His acute attack dated from March 1924. His physical state was one of mild Parkinsonism with rhythmic myoclonus in the deltoids, trapezii, and pectorals of both sides. Breathing was rapid (40 per minute) and noisy.

He left the wards of the National Hospital but continued to attend the out-patient department regularly.

When seen in August 1927 (two years and seven months afterwards), his Parkinsonism had enormously advanced, so that he was completely helpless. He was unable to make the effort of speaking. His respiratory disorder had disappeared six months previously and there were no traces of myoclonus in the shoulder muscles.

This case was reported on one or two occasions, and pneumographic records were made to show a synchrony between the rate of the myoclonic jerks and the respiration.

5. F.M., male, age 24 came into hospital in April, 1925 (Dr. Kinnier Wilson, Queen Square). An acute attack of encephalitis had occurred exactly one year before. At the time of his admission he presented the picture of Parkinsonism of moderate severity. For the past two months his breathing had become a little noisy and difficult. His chief respiratory anomaly consisted in attacks of bradypnoea in which the respiratory rate fell to 12 per minute. Thoracic expansion was of good amplitude.

In June 1927 (*i.e.*, two years and two months after first coming under observation) all traces of disordered respiration had gone. His Parkinsonism syndrome was still present and there was in fact an aggravation in some of the symptoms, especially the salivation.

6. B.A., female, age 14, first came under observation in January 1926 (Dr. Birley, at Queen Square). Her acute attack had occurred between four and five years previously. At the time of admission she complained of spasmodic, involuntary jerking movements of the head in a backward direction. This movement was accompanied by a wide opening of the mouth and a sharp, noisy inspiration. The head then slowly returned to its normal position and at the same time the mouth closed and the breath was expelled slowly and quietly. The movement occurred at two-minute intervals when she was conscious of being watched, but was less frequent when her attention was distracted. There were no gross physical signs in the nervous system beyond irregularity of the pupils and slight right facial weakness.

The chief change noted in July 1927 (18 months later) was a very definite dysarthria. The head movements and inspiratory tic had ceased.

7. W.G., female, age 17, was first seen (Prof. A. J. Hall) in September 1925. She had suffered her acute attack in February, 1924. When first seen she was definitely Parkinsonian and had periodic attacks of noisy tachypnoea. These had been present for nine months. The attacks were very frequent and lasted from three minutes to half an hour. They were accompanied by tetany, cyanosis and mental confusion; the eyes were often fixed in an attitude of extreme left lateral deviation. A period of acapnia usually supervened after each paroxysm.

She was seen again just two years afterwards; she had not had a single attack for the past year. On the other hand, her conduct had become much altered; she had developed into an incorrigible liar and thief; her Parkinsonism had not advanced.

8. J.W., male, age 16 (case of Prof. A. J. Hall) first came into hospital in March 1924, for insomnia and paroxysms of noisy tachypnoea, accompanied by tetany. The acute phase dated from 12 months previously. His physical condition was one of Parkinsonism of mild degree.

His physical state in September 1927 (three years and six months later) had not become worse, but he had now developed oculogyric crises; his conduct had also deteriorated and he had become subject to maniacal attacks with violence. The respiratory disorder had entirely disappeared.

9. L.K., female, age 9 (case of Prof. A. J. Hall), had her acute attack in March 1924, and came into hospital in December 1924, with Parkinsonism and frequent attacks of noisy tachypnoea. Her physical state in August 1927 (two years and eight months later) had become worse and she was now an inmate of a mental hospital suffering from intense depression. Her breathing attacks had left her some months ago.

10. E.W., male, age 20 (case of Prof. A. J. Hall), had his acute attack in March 1924, and three months later began to develop mild attacks of rapid breathing. In September 1925, signs of Parkinsonism were noticeable in his left arm. In September 1927 (*i.e.*, two years and three months after the onset of tachypnoea) his Parkinsonian syndrome was worse, but the attacks of rapid respiration have ceased.

11. R.W., male, age 17 (case of Prof. A. J. Hall). The acute attack dated from March 1924; In October of the same year attacks of gasping first appeared.

Six months later (by March 1925) his respiration became normal. His physical state in November 1925, was one of Parkinsonism; no further respiratory anomaly has appeared.

12. A.M., male, age 9, was admitted to the National Hospital (Dr. Kinnier Wilson) on account of nocturnal restlessness. His acute illness had occurred about 19 months previously (*i.e.*, in the Spring of 1924). Whilst in hospital his day-time conduct was exemplary, but towards nightfall he became noisy, excited, and turbulent. He would run about the ward blowing hard through his fists as though he were playing a musical instrument, and it was not until the small hours of the morning that he would drop off into a sleep of exhaustion. There were no signs of Parkinsonism. He was transferred in February 1926, to the Northern Hospital Encephalitis Unit, Winchmore Hill. Here his behaviour steadily grew worse; his blowing tic persisted up to February 1927, when it gradually left him. When seen in October, 1927 (*i.e.*, 23 months after his admission to the National Hospital) there were still no signs of Parkinsonism; his respiratory disorder had ceased; his conduct however was certainly worse than it was two years previously.

C. *Cases in which respiratory disorder has improved though not disappeared.*

Within this group we place a few cases which in the course of time have shown some improvement in the breathing attacks, either as regards frequency or severity or both. The physical concomitants may, as in both the following cases, simultaneously get worse.

13. W.E., male, age 55, first came under observation in 1924 (Dr. Phillips, Southmead Hospital, Bristol). He had been treated for acute encephalitis six weeks previously. When at Southmead Hospital there was a moderate degree of Parkinsonism, and rhythmic myoclonic movements of the left shoulder muscles. He was subject to paroxysmal attacks of noisy tachypnoea of very shallow amplitude "like the panting of a dog which has been running."

He was often in and out of hospital in the next three years. When visited in July 1927, by one of us (two years and seven months after first being seen), he was in a state of very severe Parkinsonism. He made no complaint of breathlessness or thoracic distress, but was troubled with intense vertigo. The amplitude of the chest during ordinary slight breathing was fair, but perhaps a little less than normal. When told to take a deep breath in and out, his respiration became somewhat rapid (28 per min.).

His attacks of tachypnoea still occurred though much less frequently and with less severity. They were particularly prone to come on when in bed at night.

14. E.B., male, age 31 (in-patient at Queen Square, under Dr. Howell), was admitted in January 1926. His acute attack of encephalitis dated from 1924. His respiratory disorder, which had been present five months, consisted in attacks of noisy tachypnoea in which he would stand up first of all with his trunk bent forwards to a right angle, breathing noisily and rapidly, and then suddenly straighten up and bend backwards in an opisthotonic attitude. He exhibited a mild degree of Parkinsonism.

In June 1927 (*i.e.*, 18 months after first being observed), his attacks were still present, but less severe in nature and less frequent in occurrence. He has now developed tremor in addition to his general attitude of Parkinsonism.

D. *Cases in which there has been little or no change in the severity of the respiratory disorder.*

15. H.W., male, age 26, admitted to Queen Square (Dr. Kinnier Wilson), in December 1924.

The acute attack occurred in January 1920, and was followed six months later by the gradual onset of Parkinsonism. At the time of his admission the P.A. syndrome had

reached a moderate degree of severity. He was, moreover, subject to incessant attacks of involuntary yawning in which his mouth would gape to a surprising extent. No other respiratory disorder was present, and there were no psychical changes.

His state in June 1927 (two years and seven months later), had shown no material change. The yawning bouts were as severe and as frequent as ever. The Parkinsonian state had not advanced to any appreciable extent.

16. E.F., male, age 52, was admitted in July 1926, under Dr. Collier (Queen Square). He had suffered his attack of encephalitis in December 1924. There was no trace of Parkinsonism and no psychical changes. His respiratory anomaly consisted in grotesque attacks of breath-holding, in which he stood up and took several deep, noisy breaths in rapid succession. Next he stiffened himself and slowly bent the trunk backwards, at the same time extending his arms and abducting them at the shoulder joints. The forearms and arms alternately flexed and extended. There was sometimes a transient loss of consciousness during these attacks, in which the patient would fall.

Twelve months later his paroxysms were still continuing with unchanged severity and frequency, and no fresh symptoms had appeared.

A short note on this patient appeared in a previous paper³ but was not included in the original series.

17. S.T., male, age 8, was admitted into Queen Square (Dr. Birley) in April 1924, with an acute attack of encephalitis with choreiform movements. He left hospital in the middle of May, apparently cured.

Soon after returning home he became noisy and restless at nights and developed attacks of noisy and rapid breathing, aggravated by excitement or by fatigue.

When examined in July 1927, (two years and three months later) there was no evidence of Parkinsonism, but the bouts of noisy tachypnoea were still present.

18. Male, age 14, first came under observation (Dr. A. Greig Anderson) in July 1924. His original illness had apparently occurred in June 1924. On July 1st, 1924, he was extremely restless and sleepless; no objective signs were discovered in the central nervous system except an ill-sustained nystagmus. In November of the same year he developed attacks of noisy tachypnoea, still not sleeping well; examination revealed a doubtful plantar response on the right, and nystagmus on right lateral ocular deviation. In June 1927 (*i.e.*, two years and seven months later) he was still having attacks of tachypnoea.

19. E.W., male, age 24, attended the out-patient department of Queen Square (Dr. Birley) in May 1926, for involuntary gaping movements of the lower jaw, which had been present for two months. His acute attack had occurred in June 1924; there was a condition of Parkinsonism of moderate severity. He attended the out-patient department regularly and up to the date of communication.

In September 1927 (*i.e.*, one year and four months later) there was no material change either in his Parkinsonism or in his gaping.

20. W.D., male, age 33, attended the out-patient department of Queen Square (Dr. Howell), suffering from Parkinsonism, palilalia, oculogyric crises and paroxysms of breath-holding, ushered in by a very long and audible nasal inspiration. His attack of acute encephalitis dated from January 1924. The respiratory trouble had been present for two and a half years and still continued to date (June 1927).

21. M.H., female, age 11, attends the out-patient department at Queen Square (Dr. Adie). Her encephalitis occurred in 1924; her present state is one of Parkinsonism and character changes. She worries other children, torments and assaults her sisters. For the past three years she has been subject to bouts of tachypnoea with a loud expiratory phase. There has been no change in the severity or frequency of these attacks during the last three years.

22. R.G., male, age 14, a patient of Queen Square (Dr. Holmes), whose acute attack dated from May 1924, is now in a condition of Parkinsonism. In the spring of 1925 he became addicted to a respiratory tic in which he would give a series of loud and abrupt

expirations through his nostrils. He was admitted to hospital in May 1927, and improved considerably; the tic relapsed however in July and is still present in all its former severity.

23. K.D., female, age 12, was admitted to Queen Square (Dr. Kinnier Wilson) in June 1926. She had suffered from encephalitis some time in 1924, and her present complaints were of dyspnoea and audible tachypnoea. There was a mild degree of Parkinsonism present.

In September 1927 (*i.e.*, one year and three months after leaving hospital) her physical condition had improved; except for her expressionless face there was no trace of Parkinsonism. The mother stated that she had become wilful of late and had developed a habit of stopping strangers in the street and engaging them in conversation. The respiratory disorder was still present and had not noticeably altered in character. One new feature, however, has arisen; many of her breathing attacks now end in sleep; she drops her head, and shuts her eyes for about two minutes; she is very easily aroused from this sleep.

The occurrence of these narcoleptic-like attacks following the paroxysms of tachypnoea is of especial interest. Sleep has been reported by both Mlle. Lévy and Fiamberti, coming on at the end of bouts of disordered breathing. In their patients, however, the sleep appeared to be one of exhaustion, whereas in our case, there was no preceding fatigue or background of sleepiness. The unconsciousness would come on suddenly; it was irresistible; it resembled normal sleep in character and was promptly terminated by extraneous stimuli.

24. F.H., female, age 27, attended the out-patient department of Guy's Hospital (Dr. Symonds) with a mild Parkinsonian syndrome. Her acute attack dated from December 1925. Since May 1926, she had been subject to a paroxysmal dry, hacking cough, coming on towards evening. There was no adequate cause for the cough discoverable in the chest or larynx. At times she also complained of breathlessness causing her to take every now and then a respiration of deeper amplitude. When seen in September 1927 (15 months later), the respiration disorder was still present.

25. A.H., male, age 30, an out-patient of Guy's Hospital (Dr. Symonds), sustained an attack of encephalitis some time in 1924. A year later he began to complain of numerous indefinite symptoms resembling those of neurasthenia and in addition "fits" of a vague nature made their appearance. In January 1927, he began to suffer from attacks of tachypnoea and sighing respiration particularly at meal times.

In September 1927 (nine months later), the respiratory disorder was still present, and there was no trace of Parkinsonism.

26. P.A., male, age 17, an out-patient of Queen Square (Dr. Howell), dated his original attack of encephalitis from March 1925. He slowly developed a paralysis agitans syndrome of moderate severity, and six months before coming under observation he began to have attacks of involuntary gaping. These paroxysms persisted with variable intensity, but there was no real change in their severity or frequency over a course of six months.

27. A.B., male, age 19, was admitted to the National Hospital (Dr. Holmes) for Parkinsonism. His acute attack dated from some time in 1925. For one year prior to admission his breathing had been subject to paroxysms of noisy acceleration. In September 1927 (*i.e.*, 16 months after the onset of the tachypnoea) his rapid breathing still persisted with its usual severity.

E. *Fatal Cases.*

Death may supervene both in cases of acute encephalitis with respiratory manifestations or—less commonly—in cases of chronic or subacute encephalitis in which breathing anomalies have coexisted.

28. L.B., male, age 5, was admitted to Charing Cross Hospital in January 1925, (Dr. Pearson). The acute illness had occurred in April 1924, and was followed by restlessness, insomnia, and choreiform movements. When first admitted the child was

awkward on its legs; could not talk intelligibly; it was very drowsy by day and would sometimes fall asleep over its meals. At night-time the child was intensely restless. The expression was vacant, the mouth gaping widely.

Three months later the head became retracted and Kernig's sign was present. The spinal fluid was normal. The child's respiration now became characteristically changed. It was irregular and there were periods of apnœa followed by a spell of noisy exaggerated breathing with marked expiratory phase. During the apnœic period there were simultaneous involuntary movements of the lower jaw and the left hand.

Four months after admission the child's condition had markedly deteriorated. It had become cachectic, and could swallow only with difficulty. The limbs and trunk were still more hypertonic; irregular twitching movements of the left hand were visible. The temperature rose to 103° at night. There was no change in the character of the respiration. This state lasted a month and the child eventually succumbed.

29. A.C., male, age 25 (patient of Dr. Feiling, Hospital for Epilepsy and Paralysis, Maida Vale) sustained his acute attack of encephalitis in March 1924. He thereafter developed a condition of Parkinsonism, together with paroxysms of tachypnœa with a cogwheel expiratory phase. The Parkinsonian state gradually increased and in April 1927 (*i.e.*, 37 months after the onset of the acute illness) he died. Dr. Hunt Cooke kindly wrote to us as follows: "... the irregular and peculiar respiratory condition persisted until he became comatose about a week before he died. He then had Cheyne-Stokes breathing."

Case No. 29 was among the seven cases recorded in our original paper. In this patient fluoroscopic examination of his diaphragm showed that the rise during expiration consisted of a series of rapid jerky movements. This 'cogwheel phenomenon' was also demonstrable in the pneumographic tracings.

We have purposely omitted from this section any record of fatal acute cases in which the terminal phase was characterized by acceleration of the respiration rate. A consideration of such does not come within the scope of this present paper.

DISCUSSION.

In all, some 29 cases have been followed up over periods extending to three years; the results have been variable and may be set out thus:—

A.—Cured	2
B.—Respiratory disorder cured but other manifestations worse or unchanged	10
C.—Respiratory disorder improved but other manifestations worse or unchanged	2
D.—Respiratory disorder unchanged	13
E.—Death	2

One patient could not be traced.

It appears therefore that the respiratory disorder disappeared in 12 out of 29 cases and improved in two others: in the remaining 15 cases it has persisted unchanged up to time of observation and in two instances till death.

(a) *The variety of respiratory disorders.*

In Table 1 the results are set out according to the variety of the original breathing disorder. Tachypnœa was the commonest variety, comprising 15 cases. Of these, nine were cured (as far as the respiration was concerned) when followed up over periods extending from 27 to 42 months: one patient

had died. The remaining six cases remained unchanged when seen at a period of nine months to three years later. Spells of breath-holding constituted the next commonest type, five cases belonging to this group; of these patients, one was cured, one had improved, and one died; the other two had improved. Of the three cases of paroxysmal yawning, all remained *in statu quo* after intervals from six months to over two and a half years. The other cases of tics (coughing, blowing, etc.), numbered four in all, two of which remained as before and two improved. One patient only was observed with bradypnoea. In this case the respiration became normal although the Parkinsonism increased.

(b) *Influence of the Parkinsonian Syndrome.*

The effect of an associated state of Parkinsonism is shown in Table II. In 20 cases, the patient was also afflicted with some degree of the paralysis agitans syndrome. Of these, one patient died. In eight, the respiratory disorder was unaltered; in two it improved and in nine it disappeared. The fate of the breathing anomaly bore no parallelism with the severity of the syndrome or with its course.

The nine cases in which no Parkinsonism coexisted included one fatal case, three cures, and five in which the respiration did not change.

(c) *The presence of psychotic manifestations.*

The possible influence of concomitant disorders of conduct or character upon the prognosis of respiratory disorder offers a suggestive avenue of enquiry, and may have a bearing upon the causation of the abnormal breathing. Psychotic changes of a gross character were present in eight only of this series; of these, one became well, and in five the respiration became normal although the other manifestations did not improve. In the other two instances the respiration remained unchanged (see Table III).

Of the remaining 21 patients in whom there was no obvious behaviour or character change, two died, six became well, two improved, and eleven remained *in statu quo*.

(d) *The age factor.*

Fifteen of our patients were in the second decade of life; three were in the first, and six in the third; two patients were in the fourth and fifth respectively and one only in the sixth. Males outnumbered females in our series, the proportion being twenty-one to eight.

(e) *The date of the original attack.*

Although the literature shows that respiratory disorders may follow encephalitis whichever epidemic the original illness dates from, it is the experience of most in this country that the 1924 epidemic was responsible for the majority of the cases of postencephalitic respiratory disorders which are seen to-day. This impression is supported in our series; Table IV indicates that no less than 23 out of 29 cases date the original illness from the 1924 epidemic.

Reference to official figures, however, indicates that a proclivity of the 1924 cases to develop respiratory disorders is apparent rather than real, for the epidemic of that year was the severest known in this country. The number of notifications is almost double those occurring in the following year, which marked the next most severe epidemic.

THE VARIABILITY OF THE RESPIRATORY DISORDERS.

Postencephalitic respiratory disorders are in most instances paroxysmal in appearance, and it is of value to consider those factors which appear to influence the severity or frequency of the attacks. In the majority of cases no precipitating cause can be traced; their proximity is heralded at times by prodromata of thoracic, laryngeal or nasal discomfort, and the disordered breathing follows. Frequently it is noticeable that the attacks occur only towards nightfall, and in such there seems to be a close association with fatigue states; the converse condition, however, in which the subject is at his worst in the early hours of the morning, shortly after waking, is not uncommon. In a few patients the respiratory disorder invariably follows the awakening from sleep, irrespective of the hour. Occasionally (Case 3) the respiratory anomaly occurs only during sleep, and the patient is ignorant of any abnormality.

Emotional stress is perhaps the most potent aggravating factor in these cases; almost all these patients are at their worst when the object of scrutiny, or when under the influence of fright, annoyance, or embarrassment. Pleasurable excitement, however, may be accompanied by a respite from the attacks, as in Cases 14, 16, and 23. Boredom or lack of interest in the environment is usually associated with an aggravation; thus we are told by the wife of patient No. 14: "He is better when occupied; he can go out for a five mile walk or a motor ride and not have an attack; but the trip must be one of interest to him; he must *want* to go out." If on the other hand he is reluctantly led out by his wife for a stroll the attacks are frequent. During concentration, or occupations entailing mental or physical effort, there may be a diminution in the paroxysms; thus, one patient (Case 14) was fond of music and would like to compose and play pianoforte music for an hour at a stretch, during which time he would be free. The act of shaving however was always attended by unpleasant associations and he was invariably worse whilst so occupied.

The effect of intercurrent illness or noxious stimuli in diverting the respiratory disorder is noteworthy. In Case 1, for example the patient was almost free from attacks whilst suffering from pneumonia. We are informed that another patient (Case 14) was once taken out for a walk against his will; instead of the attacks being worse as was anticipated his breathing was normal because "he had very bad chilblains on his feet and these hurt him intensely as he walked." A temporary freedom coinciding with a feverish cold has been noted in more than one case.

The act of eating has been known to exert an unfavourable influence in a few cases; a patient (Case 15) tells us: "It all depends upon the amount of

phlegm I have in my mouth ; when I have a lot my mouth opens. . . . It is always worse when I am eating or when my food is lying heavy on my chest."

Severe physical exertion as a rule has little or no influence upon the appearance of respiratory disorders ; alterations of posture are occasionally possible factors in curtailing or launching an attack.

The ability to cut short an attack is rarely present to any marked extent on the part of the patient himself. At times we hear that by exerting a very determined effort of will the respiratory disorder may be checked, but usually for a while only. The effort of self-control apparently entails so much distress that the patient abandons it and finds it easier to endure the paroxysm. Extraneous stimuli are more potent ; by speaking sharply to the patient, or by distracting the attention, the breathing disorder may sometimes be checked. More often it is possible to correct the respiration by directing the patient's attention towards his own performance and by ordering him to hold his breath or to take deep, slow inspirations. The effect however is rarely more than temporary, and becomes less efficacious on repetition.

In no case in our experience has any permanent benefit resulted from such measures as heavy dosage with sedatives, from electrotherapy, or from the inhalation of oxygen, nitrous oxide, or carbon dioxide. Of the cases in the present series which cleared up, no association could be demonstrated with any particular therapeutic measure ; the majority were actually on no treatment at all.

There is an obvious similarity therefore between the results of precipitating and inhibiting factors and the phenomenology of hysteria, but the association must not be strained. The power of temporary inhibition of disordered breathing at the instigation of external suggestions argues merely that the volitional or corticobulbar controlling mechanism is intact ; the aggravation of the attacks during emotional stress and during introspection illustrates a phenomenon occurring in every type of organic disease in which consciousness is intact.

It seems therefore that though diurnal variability is the rule—dependent upon the play of internal and external factors—the influence of such stimuli is but slight in degree and transitory.

Occasionally the respiratory disorder changes in type ; thus a particular patient may show at the onset of his illness a paroxysmal cough which later is replaced by paroxysms of tachypnœa, or of breath holding. Such was the case with Nos. 1 and 23 in our series (cf. also Wimmer's case 12).

SUMMARY.

Although the number of cases which form the basis of this paper is too small to justify definitive conclusions as to prognosis, it is possible that these cases constitute a representative example of postencephalitic respiratory disorders in general. In the 29 cases investigated, almost every variety of known breathing anomaly is represented ; in general terms it may be said that nearly

one half of the cases have either ceased to show any respiratory disorders, or else have shown them to an attenuated extent. An equal proportion of cases remained unchanged over a period of time, extending up to three years. Two patients have died, whilst still giving evidence of a respiratory disorder.

It would seem that the character of the breathing anomaly does not materially alter the prognosis, although the small series of cases investigated will not allow of any conclusive statement. The proportion of recovery cases appears to be greater in the cases with a progressive Parkinsonian state than in those without the manifestation of paralysis agitans.

Although it is indubitable that numerous physical and psychical stimuli may temporarily influence the severity of the disorder, there is no evidence that these factors are other than temporary.

It must be emphasized that the recovery rate apparently does not depend upon any particular treatment; improvement when it occurs seems to appear spontaneously.

We beg to thank the Physicians of the National Hospital for permission to quote many of the cases in this series.

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TABLE I.

THE OUTCOME OF THE RESPIRATORY DISORDERS CORRELATED WITH THE CHARACTER OF THE BREATHING ANOMALY.

(The letters A, B, etc., refer to the groups considered in the text.)

Variety of Disorder.	Case No.	Time (in months) over which observed.				
		A	B	C	D	E
TACHYPNEA	2	32	—	—	—	—
	3	—	32	—	—	—
	4	—	31	—	—	—
	7	—	24	—	—	—
	8	—	42	—	—	—
	9	—	32	—	—	—
	10	—	27	—	—	—
	11	—	35	—	—	—
	13	—	—	31	—	—
	17	—	—	—	27	—
	18	—	—	—	31	—
	21	—	—	—	36	—
	23	—	—	—	15	—
	25	—	—	—	9	—
	27	—	—	—	16	—
	29	—	—	—	—	37
BRADYPNEA	5	—	31	—	—	—
BREATH-HOLDING	1	36	—	—	—	—
	14	—	—	18	—	—
	16	—	—	—	12	—
	20	—	—	—	30	—
	28	—	—	—	—	5
YAWNING	15	—	—	—	31	—
	19	—	—	—	16	—
	26	—	—	—	6	—
BLOWING, SNIFFING	6	—	18	—	—	—
	12	—	23	—	—	—
	22	—	—	—	30	—
COUGHING	24	—	—	—	15	—

TABLE II.

THE PARKINSONIAN SYNDROME AND ITS ASSOCIATION WITH RESPIRATORY DISORDERS.

		Time (in Months) over which observed.					
		A	B	C	D	E	
WITH PARKINSONISM.	MILD	—	18	—	—	—	
		—	—	—	15	—	
	MODERATE	—	42	—	—	—	
		—	—	—	5	—	
		—	32	—	—	—	
		—	26	—	—	—	
		—	24	—	—	—	
		—	32	—	—	—	
		—	27	—	—	—	
		—	35	—	—	—	
		—	—	18	—	—	
		—	—	—	36	—	
		—	—	—	39	—	
		—	—	—	16	—	
		—	—	—	31	—	
		—	—	—	6	—	
		SEVERE	—	—	—	16	—
			—	—	—	—	37
	—		—	31	—	—	
	—		31	—	—	—	
TOTAL		—	9	2	8	1	
WITHOUT PARKINSONISM	36	—	—	—	—		
	32	—	—	—	—		
	—	23	—	12	—		
	—	—	—	27	—		
	—	—	—	31	—		
	—	—	—	30	—		
	—	—	—	9	—		
	—	—	—	—	5		
TOTAL		2	1	—	5	1	

TABLE III.

CORRELATION OF RESPIRATORY WITH BEHAVIOUR DISORDERS.

Type of Case.	Time (in Months) over which observed.				
	A	B	C	D	E
WITH CONDUCT OR PSYCHOTIC CHANGES.	36	32	—	15	—
	—	24	—	36	—
	—	42	—	—	—
	—	32	—	—	—
	—	23	—	—	—
TOTAL	1	5	—	2	—
WITHOUT CONDUCT OR PSYCHOTIC CHANGES	32	31	31	31	5
	—	26	18	12	37
	—	18	—	27	—
	—	27	—	31	—
	—	35	—	16	—
	—	—	—	30	—
	—	—	—	30	—
	—	—	—	15	—
	—	—	—	9	—
	—	—	—	16	—
TOTAL	1	5	2	11	2

TABLE IV.

SHOWING THE YEAR OF THE ORIGINAL ATTACK IN THE PRESENT SERIES OF CASES. OFFICIAL STATISTICS ARE ALSO APPENDED, GIVING THE TOTAL NUMBER OF NOTIFICATIONS AND DEATHS OCCURRING IN ENGLAND AND WALES.

Date of Original Illness.	NUMBER OF CASES.						Figures for England & Wales.	
	A	B	C	D	E	Total.	Notifica-tions.	Deaths.
1919 ..	—	—	—	—	—	0	541	294
1920 ..	—	—	—	1	—	1	890	480
1921 ..	—	—	—	—	—	0	1,470	729
1922 ..	—	1	—	—	—	1	454	339
1923 ..	—	1	—	—	—	1	1,025	531
1924 ..	2	8	2	9	2	23	5,039	1,407
1925 ..	—	—	—	3	—	3	2,635	1,372
1926 ..	—	—	—	—	—	0	2,267	1,324