FACIAL IRRITABILITY: ITS RELATION TO CONVULSIVE DISORDERS, AND ITS GENERAL CLINICAL SIGNIFICANCE, IN INFANCY, AND ALSO IN LATER CHILDHOOD.

By CHARLES M'NEIL, M.A., M.D., F.R.C.P.E., Lecturer on Diseases of Children, University of Edinburgh.

The sign of facial irritability, also commonly known as Chvostek's sign, is generally accepted as a clinical indication of recent, present, or impending convulsive disorders of a particular kind in infants. These convulsive seizures are of three kinds, attacks of laryngeal spasm or laryngismus stridulus, general convulsions, and the peculiar cramp-like spasm of the hands and feet known as tetany: and one or two, or all of these may occur together. Other kinds of spasmodic seizure are also alleged to occur. But those mentioned are agreed to form an associated group and have received the group name spasmophilia. Spasmophilia is then that condition of body in children of tender age (the great majority of the cases are between six months and two years of age) in which definite types of convulsive seizure have occurred or are likely to occur.

There is practical agreement that, when the sign of facial irritability is present in an infant, the condition of spasmophilia exists, and that one or more of these types of convulsive seizure is actually present, or is likely to occur. Facial irritability is accepted as the sign of manifest or of latent spasmophilia. But it has also been known for many years, that facial irritability occurs after the age period when spasmophilia is met with. Its incidence and its significance in older children is, however, the subject of dispute.

In the following paper, the results of the test of facial irritability in 403 cases are analysed. For the two periods of infancy (terminating at age 2½ years) and childhood up to 13 years, an answer has been sought for two questions. What is the association of facial irritability with muscular convulsions of any kind? What is the general morbid and clinical significance of a positive test?

The Test.—Some remarks may be made about the performance of the test; its variations and fallacies. Facial irritability is frequently known as the facial phenomenon: but the former word is more expressive of the condition indicated—an unusual
degree of irritability of the superficial fibres of the facial nerve in the face, so that a light, sharp tap of the finger on the cheek excites a lateral twitch of the middle of the upper lip. In a higher degree of irritability, there is also visible a muscular twitch in the side of the nose, and in the forehead above the root of the nose. The best position for this mechanical stimulus is midway between the angle of the mouth and the angle of the jaw. As a rule, if the reaction is present on one side, it is also and equally present on the other.

Facial irritability of this kind is not an isolated phenomenon: it is part of a general increased irritability of peripheral nerves. And evidence of this general irritability may be found elsewhere; in the arm by a similar light, sharp percussion of the musculo-spiral nerve in the upper arm; of the posterior interosseous nerve on the back of the elbow; and in the leg by tapping the exposed peroneus nerve as it winds round and below the upper end of the fibula. On the whole, it is easier and more convenient to make the test on the face, but in restless and crying infants it may be impossible at the time to obtain the sign on the lip, and quite easy to do so in the leg. The following example will illustrate this:

John S., aged 1½, microcephaly, with frequent laryngismus; no signs of rickets; the baby always restless and bad-tempered. Facial irritability repeatedly tested, was once doubtfully positive. Leg irritability tested at the same time, was always unmistakably positive.

In the present investigation there were a few cases of this kind, the sign of irritability being absent in the face, but present in the leg or arm. These cases have been included in the following tables of positive facial irritability. Hereafter facial irritability will be denoted by the abbreviation F.I.

When we speak of irritability of a peripheral nerve to mechanical stimulation, we perhaps assume too readily that the irritability is in the nerve. For it is possible that it may be an increased contractility or irritability of muscle that we are dealing with. This possibility of a muscle irritability may be alluded to later in the clinical discussion.

The electrical excitability on the opening or closing of measured galvanic currents (Erb's and Thiemich's signs) probably tests the same quality of the neuro-muscular apparatus. It seems to be a more exact test than that of F.I.; it has not been shown to be more delicate; it is certainly not nearly so convenient. It was not employed in the present investigation.
Facial Irritability

**Fallacies of the Test.**—Certain apparent positive reactions should be excluded. In fat, healthy babies, the elasticity of the tissues of the cheek allows the propagation to nose and lip of a percussion wave: but this is quite different from the lateral twitch in the centre of the upper lip and in the side of the nose of a true F.I. Dr John Thomson has also warned against mistaking for F.I. the slow, deliberate pursing of the lips in the lip reflex of young babies. Further, it is not safe to accept a twitch of the eyelids as a positive test.

**Variations.**—Pseudo-negative reactions are obtained when the child is crying. At such times, leg irritability (the so-called Peroneus Phenomenon of Lust) should be tested. Also for an hour or two after general convulsions, F.I. is in abeyance. I have repeatedly tested for it immediately after general convulsions have ceased, and have never yet obtained a positive F.I. at this time. But in one case it was again present after three hours, and frequently was observed to return in the same day.

Some of these variations in the test deserve special mention. Thus Mosse has described the disappearance of the test in three cases where fever developed. I can confirm this from my own observations, having watched the sign disappear during the onset of febrile bronchitis in rachitic babies, and return when the feverish attack was over. Anderson and Graham have also observed the disappearance of the test during fasting. These variations have been attributed to acidosis. On the other hand, I have seen marked F.I. first develop in the course of high fever. There is also considerable variation in the degree of the reaction; and it is of practical importance to separate an ordinary or slight reaction, where only a localised twitch in the middle of the upper lip or nose takes place, from a severe reaction where there is simultaneous and even repeated twitching of the lip, nose, and lower part of the forehead. These severe reactions are more common in infancy than in later childhood. On the other hand, a positive reaction is more likely to come and go in an infant than in an older child.

**General Results.**—The table on next page shows the general results obtained.

I have not attempted to give percentages of incidence. Besides the too small numbers investigated, there would be other fallacies in such a statement. But the numbers dealt with at least give a considerable number of positive reactions.
for analysis; and also show that positive reactions are by no means rare after the age period of spasmophilia.

**TABLE I.**

| Age Period                | Total Number | F.I. present |
|---------------------------|--------------|--------------|
| Birth to 5 months         | 47           | 4            |
| 6 months to 21/2 years    | 169          | 34           |
| 21/2 years to 41/2 years  | 79           | 15           |
| 5 years to 13 years       | 108          | 22           |
| Birth to 21/2 years       | 216          | 38           |
| 21/2 years to 13 years    | 187          | 37           |
| Total                     | 403          | 75           |

Dividing the whole group into two at the age of 21/2 years, the first period should include the great majority of cases of spasmophilia, while the later period should contain different clinical conditions.

Table II. gives the analysis of positive reactions in the period up till 21/2 years, which may also be called the period of spasmophilia.

**TABLE II.**

*Positive F.I. Birth to 21/2 Years.*

| Clinical Condition                      | Positive F.I. |
|-----------------------------------------|---------------|
| Spasmophilia, with or without rickets   | 24            |
| Rickets without spasmophilia           | 6             |
| Neither rickets nor spasmophilia       | 8             |
| Total                                  | 38            |

The majority of positive F.I.s during this period of early life is thus made up of cases of spasmophilia and rickets. Before examining further these two groups, it will be convenient to deal with the third group of 8 positive F.I.s in Table II., which were neither rickets nor spasmophilia. These included 2 cases of bronchitis in which the presence or absence of rickets was not noted; 2 cases of tuberculous meningitis with convulsions, in which F.I. developed at a late stage in the illness.
Facial Irritability

and again disappeared before death; 2 cases of convulsions in infants, aged six and seven weeks, one being syphilitic, and the other idiopathic convulsions, and in both of which leg irritability alone was present; a case of pneumonia in which F.I. developed intensely during high fever and was accompanied by fine tremor of the face and hands; and an apparently healthy breast-fed infant of seven months with two teeth. Of these 8 cases, it may be said of them, that either convulsions were present, or that in the remainder it would be difficult to exclude a slight degree of rickets. At least, if a strong association of rickets and spasmophilia with facial irritability can be shown on other grounds, it need not be weakened by the existence of this small number of cases apparently distinct from these conditions.

It may be noted that F.I. was negative in two cases of congenital laryngeal stridor.

Spasmophilia, Rickets, Facial Irritability.—The mutual relations of these three conditions, as shown by this investigation, can be given in a tabular way as follows (Table III.):

|                      | Total | Positive F.I. | Negative F.I. |
|----------------------|-------|---------------|---------------|
| Spasmophilia, total  | 26    | 24            | 2             |
| with rickets         | 23    | 21            | 2             |
| without rickets      | 3     | 3             | 0             |
| Rickets without spasmophilia | 32   | 6             | 26            |

The following conclusions seem warranted by these figures:

1. The great majority of cases of spasmophilia show the sign of facial irritability.
2. The great majority of cases of spasmophilia are rachitic.
3. Only a small number of cases of rickets without spasmophilia show facial irritability.

A further point of practical importance may be raised. If a case of rickets, which up to the present has had no convulsions local or general, shows a positive F.I., have we any guide as to the likelihood of such convulsions taking place? I would suggest that a prognosis of spasmophilia in such cases may be assisted by attention to two things; the severity of the
Charles M'Neil

facial sign; the softness and lack of tone of the muscles. We have already seen that nearly all spasmophilic infants show some degree of bony rickets; but the extent of the bony lesions does not always correspond with the severity and frequency of the convulsive disorders. In my experience there is a closer correspondence between the softness and flabbiness of the muscles and ligaments, and the severity of the fits and laryngismus.

General Convulsions with Negative Facial Irritability.—In the group under age 2½ years, there were 10 cases of general convulsions, not apparently spasmophilic nor rachitic, and independent of any known irritant or infection. In one of these, an infant of seven weeks, with frequent recurring fits apparently of idiopathic type, F.I. was positive. In all the others, F.I. was negative. This suggests that general convulsions may occur in this period of life independently of that irritability of the peripheral nerves which is present in nearly all cases of the group of convulsive disorders called spasmophilia.

Facial Irritability after 2½ Years of Age.—It has already been said that there is general agreement as to the significance of Chvostek's sign of facial irritability in infancy. It indicates actual, or impending convulsive disorders, of a special kind called spasmophilia.

The occurrence of F.I. in older children has long been known. But its interpretation at this later age has been and remains a matter of dispute. Some, and notably Hochsinger, have declared that a positive F.I. after infancy has always a definite pathological significance, though a very different one. Hochsinger definitely teaches that a positive F.I. is always the mark of a neuropathic constitution which is congenital, and very often hereditary; and which shows itself in general nervous irritability or in some specific neuroses such as pavor nocturnus, migraine, nervous anorexia, enuresis, masturbation, etc. Raudnitz, on the other hand, found the sign more and more frequently present in children over 5 years of age, and declared that it had no pathological significance after this period.

In my series of cases, F.I. was present in 37 cases over the age of 2½ years, and 22 of these were over 5 years. The clinical grouping of most of these cases of positive F.I. is shown in Table IV.
Facial Irritability

TABLE IV.

| Clinical Condition                                      | Total Number | Positive F.I. | Negative F.I. |
|--------------------------------------------------------|--------------|---------------|---------------|
| General recurrent convulsions over 2½ years             | 13           | 3             | 10            |
| Asthma over 2½ years                                    | 9            | 0             | 9             |
| Meningitis, cerebro-spinal                              | 4            | 1             | 3             |
| Tuberculous                                             | 10           | 2             | 8             |
| Encephalitis, epidemic                                  | 10           | 0             | 10            |
| Spastic diplegia (Little's disease)                     | 5            | 0             | 5             |
| Chorea                                                  | 8            | 4             | 4             |
| Recurrent, or cyclical vomiting                         | 6            | 2             | 4             |
| "Nervousness," with intestinal worms of various kinds   | 15           | 4             | 11            |
| Coeliac disease                                         | 19           | 7             | 12            |
|                                                        | 8            | 5             | 3             |

General Convulsions.—In this condition, at the later age period, only a small number of the total showed positive F.I. And in this small group, three in number, one is not justified in assuming that the condition of irritability of the peripheral nerves had a definite association with the fits. Two of these cases may be briefly cited.

M. M., aged 10½, onset of fits, age 8 years, occurring about once a month, and nearly always at night; duration of fits about one minute. Two fits occurred in hospital in January 1923. They became less frequent, and the last occurred in October 1923. Seen in September 1924, no further fits having occurred. F.I. tested repeatedly from December 1922 to September 1924 was always positive. There was some chronic disorder of digestion with disturbed sleep (he walked in his sleep while in hospital).

Harry G., aged 8. Frequent daily fits resembling petit mal for a year. While in hospital, four slight but definite attacks of petit mal in nine days (July 1924). After discharge from hospital, fits have continued unchanged. F.I. positive in hospital, and also when seen again in October 1924.

In these two cases, F.I. was positive and remained so in both, although in one the fits ceased, and in the other they continued. The relation of facial irritability to the fits is therefore obscure.

And in the majority of recurrent convulsions of later childhood (they may be called epilepsy) F.I. is absent.

Asthma.—F.I. was absent in all cases of asthma tested. This definite result has a negative value. For in the period of infancy, the convulsive spasm of the larynx, known as laryngismus, is closely associated with facial irritability; but in later
childhood, asthma seems to occur quite independently of this condition.

Organic Cerebral Disease. — Under this group, I have arranged cases of meningitis, of encephalitis, and of old cortical hemorrhage. Here F.I. was definitely negative, except in three cases of meningitis in which it developed at an advanced stage of the disease. In the cases of epidemic encephalitis, and spastic diplegia, F.I. was absent in all. So that we may say that definite organic disease of the meninges and of the brain, producing definite symptoms of cerebral irritation, does not produce facial irritability.

Chorea. — Out of 8 cases, 4 showed positive F.I. In all of these, the reaction was slight, and in three was only temporary and soon disappeared. It seemed to have no relation to the severity of the disease, and in acute cases of the disease it was definitely absent.

Recurrent and Cyclical Vomiting. — Of 6 cases, 2 showed positive F.I.; but one of these had associated tetany, and with the disappearance of the tetany F.I. disappeared. So that no connection can be drawn between facial irritability and this condition.

Nervousness. — I have used this vague term “nervousness” in order to express the vagueness and obscurity of the condition which it denotes. Hochsinger, in speaking of a “neuropathic constitution,” includes under this pseudo-scientific and pseudo-definite term, a number of conditions in which there is a general or local disorder of the nervous system; and states that facial irritability in and after infancy is a sure mark or stigma of this neuropathic constitution.

In the group, “nervousness” with intestinal worms, there were 4 cases of positive F.I. out of a total of 15; that is to say, in 11 cases of the condition, with varying symptoms of functional nervous disorder, chiefly irritability of temper and disturbed sleep, F.I. was negative. That seems sufficient to disprove any definite connection between facial irritability and the nervous symptoms produced by intestinal worms. Some other explanation must be found in the cases where F.I. was positive; all that can be said is that in a few cases of intestinal worms, F.I. occurs.

The second sub-group, “nervousness” of various kinds, were a very miscellaneous collection of children; they included cases of habit spasm and motor restlessness, irritable or tearful
Facial Irritability

children generally with signs of chronic dyspepsia, cases of enuresis, masturbation, pica, hysterical vomiting. This group yielded 7 cases of positive F.I. out of a total of 19. But again, the important result for this, as for the other sub-group, is that a majority of cases of this kind with functional nervous disorder show no facial irritability. An explanation of the positive reaction in the other cases will have to be sought outside the condition, “nervousness” or “neuropathic constitution.” The results obtained in both these groups lend no support to the teaching of Hochsinger.

Coeliac Disease.—The results in this group are worth careful attention. The diagnosis of coeliac disease is sometimes difficult. I have included in this group 2 cases where the diagnosis was uncertain on clinical grounds, and in both of these F.I. was negative.

I have excluded 3 cases of children under 2½ years where rickets was also present, but where the condition of the stools was typical of coeliac disease; in all of these F.I. was positive. Even with inclusion of doubtful cases, a majority of the cases, 5 out of 8, showed a positive F.I. Of the 3 negative F.I.s, 2 were doubtful cases, and the third, though a definite case of coeliac disease, was only tested on one occasion.

Of the 5 positive cases, 2 showed also signs of spasmophilia, in one case tetany, in the other recurring convulsions of mild type. The facial reaction was definite in all cases, but in a majority disappeared with improvement of the condition. One clinical example from this group may be given.

Maisie F., aged 6½, admitted 9.5.24. History of loose motions for a year, and of cramps in the hands and feet for one month. On admission, the child was small, not emaciated; typical large pale pultaceous stools with excess of fat; hands and feet showed characteristic tetany. F.I. positive. Tetany disappeared in a few days, but F.I. remained, although diminishing, and even disappearing in periods of improvement. Discharged from hospital 22.8.24. Seen again 30.10.24, no return of tetany, but general condition and type of stool still that of moderate coeliac disease; F.I. present to a severe degree (simultaneous twitch of lip, nose, and forehead).

The present series of cases of coeliac disease is small, and does not justify too definite conclusions. But let us assume for the moment that the occurrence of facial irritability is a
characteristic feature of the disease, and attempt to find an explanation. The pathogenesis of coeliac disease is obscure, but one of its definite results is delayed ossification of the bones. There is also excess of calcium in the stools in the form of fatty soaps. There is thus in coeliac disease a disturbed calcium metabolism; a deficient intake; and a defective distribution to the bones, and it may be also to the nervous and muscular tissues. The increased irritability of nervous and muscular tissues deficient in calcium has a definite experimental basis.

Some of the links in this chain of argument are in the meantime hypothetical, but as an hypothesis the suggestion is put forward that the frequent occurrence of facial irritability in the present series of cases of coeliac disease is due to a disturbed calcium metabolism and to a deficient supply of calcium to the nerves and muscles.

It is obvious that the same explanation of a disordered calcium metabolism can be made for those cases of rickets, and of spasmophilia associated with rickets, that show positive facial irritability. The evidence is here more complete, for in a majority of cases of active spasmophilia, a defect of calcium in the circulating blood can be demonstrated. The nature of the calcium disturbance in rickets is an obscure problem: there need be no defect in the calcium supply, no deficiency in the circulating blood; but the defect of lime salts in the bones in definite rickets is beyond dispute. But it should be noted that facial irritability does not occur in nearly all cases of rickets. In the present investigation, there were 56 cases of bony rickets, and only in 28 was facial irritability present.

Table IV. does not give the complete list of cases of positive F.I. after 2½ years of age. The remainder could not be definitely grouped; the majority were cases of chronic dyspepsia; there was also a case of juvenile dementia, a case of convalescence after pneumonia, and so on. There seemed no clue to the explanation of the occurrence of facial irritability in this scrap-heap group. The following is an account of one of these cases:—

M. M., aged 4½, admitted 21.11.22 in a fit. Recovery of consciousness after some hours, when F. I. was strongly positive, and remained so for six weeks. Peculiar nervous condition, with fine tremors of face and hands. Cerebro-spinal fluid withdrawn under great pressure, but without increase of cells or albumen. Steady
Facial Irritability

improvement in general and nervous condition. F. I. negative on discharge from hospital. Seen again on 14.12.23, age 5½; general health good; at times is cross; also grinds teeth at night and is restless in sleep. F. I. strongly positive.

In the above case the relation of the facial irritability to the original fit, coma, and nervous tremor is quite obscure; and there is no real clue to its interpretation. The condition of mild dyspepsia with disturbed sleep should be noted; it is frequently present in cases of facial irritability in older children.

General Conclusions.

The principal results of this investigation may be stated as follows:—
1. The clinical significance of facial irritability is very different in infancy (up to 2½ years), and in later childhood.

Infancy.

2. In infancy it is definitely associated with the convulsive disorders known as spasmophilia and including laryngismus, general convulsions, and tetany.
3. At this period of life, facial irritability indicates that spasmophilic convulsions (one or more kinds) have recently occurred, or may occur in the future.
4. The likelihood of return of active spasmophilia may be judged by the severity of the facial reaction, and by the degree of muscular atony present.
5. In the great majority of cases of spasmophilia bony rickets is also present.
6. The severity of the bony lesions of rickets is an unreliable guide to the danger of active spasmophilia.
7. In one-half of 56 cases of bony rickets, facial irritability was absent.

Later Childhood.

8. Over the age of 2½ years there is no apparent association of facial irritability with general convulsions, or with asthma. The great majority of cases of general convulsions at this period occurs without increased irritability of the peripheral nerves to mechanical stimulation.
9. At this period, facial irritability does not indicate that general convulsions have occurred or are likely to occur.
10. Facial irritability does not indicate a neuropathic...
Charles M'Neil

constitution, or any recognised functional disorder or organic disease of the nervous system.

11. In most cases of older children, the sign has no definite nor serious pathological significance.

12. It is met with most frequently in cases of disordered digestion, mild and severe.

13. Its frequent occurrence in cases of cœliac disease suggests that it may be the result of disordered calcium metabolism in that disease, and in other conditions of chronic dyspepsia.

14. Its still more frequent occurrence in cases of rickets complicated with spasmophilia may be due to the same condition of disordered calcium metabolism.

References.—G. H. Anderson and S. Graham, Quart. Journ. Med., xix., p. 62. K. Hochsinger, Wien. klin. Woch., 1911, xxiv., p. 1487. Mosse, Jahrb. f. Kinderheilk, 1922, xcix. p. 244. John Thomson, Clinical Study of Sick Children, 1921, p. 438.