Remarks

ON

SOME FORMS OF SPASM IN CHILDHOOD:
THEIR CAUSE AND TREATMENT*

BY

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It has been said¹ that "a child is the sum total of his movements or tendencies to move." Movement follows on muscular contraction resulting from nervous stimulation, and spasm in ordinary medical parlance is a term used to mean involuntary sustained muscular contraction occurring as an abnormal manifestation. Such spasm may be usefully considered as it discloses itself to the clinician at different sites and in different bodily conditions.

An abnormal condition of neuro-muscular apparatus—for example, spasm—may be produced by a local peripheral cause, or result from a primary lesion or abnormality of the central nervous system interfering with its function of nervous control. It is not possible, however, to dissociate the mental functions and the functions of nervous control in the central nervous system, and spasm may occur from disturbance of the mental functions, with no detectable abnormality in local neuro-muscular apparatus or in the central seat of nervous control. Thus emotional disturbance may be the primary exciting cause. Careful attention must also be given to the degree of development of the intellectual powers in childhood. If, then, we have to take into account such varying factors in etiology, we must go further and consider important influences affecting the functions of the central nervous system. Such, in this connexion, are the influence on bodily processes of endocrine secretion, calcium metabolism, and the phenomena of anaphylaxis; the results of training and education; and the development of inhibition.

The symptom of spasm may be considered in four groups.

1. Congenital in origin, associated with local organic change in infancy, and with no detectable lesion of the nervous system.
2. The result of gross intracranial abnormality or lesion, congenital or acquired.
3. A symptom of disease resulting from vitamin deficiency in infancy.
4. A symptom dependent on nervous hyperexcitability, congenital or acquired, temporary or permanent.

Consideration here will be restricted to some conditions in Group 4. While these occur largely in "nervous" children, there is an interesting aspect of abnormal nervous excitability which is capable of practical demonstration—namely, the phenomenon of facial irritability.

CHVOSTEK'S SIGN

It has long been well known that a positive Chvostek's sign (facial irritability) is no uncommon phenomenon among children of school age, both healthy and debilitated. Eckert² investigated the frequency of its occurrence in 13,854 Austrian children and found it present in (1) 22.8 per cent. of boys and 20.1 per cent. of girls in Vienna; (2) 7.1 per cent. of boys and 7.8 per cent. of girls in provincial cities; and (3) in an intermediate percentage in the country districts. Various observers^{3,4} in this country have also found that it is by no means rare, but its significance in children of school age is disputed. As a result of the examination of

a consecutive series of 400 children between the ages of 6 and 14 I found it present in twenty-eight, or 7 per cent.

It is impossible to believe that the sign has no significance. In spite of its obscurity, there seems to be a type of child in which its presence points to certain tendencies in reaction to a specific treatment. For some considerable time I have given to practically all asthmatic children, after treatment in the ward, a prolonged course of treatment by ephedrine supervised in the out-patient department. This has usually had good results, and never untoward ones, except in five cases in which it was stated that the children suffered some "collapse," lethargy, tendency to faint, shortly after taking the medicine. In these five cases F.I. was marked, and in only two, in which F.I. was negative, have symptoms of a minor degree occurred. This will be referred to later when discussing asthma; at present it is sufficient to say that the reaction appears to depend upon the degree of alteration in systolic blood pressure brought about by the administration of a particular internal secretion in a child of unstable nervous constitution.

BLOOD CALCIUM CONTENT IN SPASMODIC CONDITIONS

The presence or absence of the sign frequently varies much from time to time in the same child, but it is often present with a normal blood calcium content. In cases of tetany in infants it has been shown⁵ that the calcium in the blood serum is greatly reduced. Italian investigators have found that a solution of calcium when applied to the brain diminishes its irritability to electrical stimulation,⁶ while the peripheral nerves are even more sensitive to lack of calcium than the central nervous system.⁷ It is interesting, therefore, to consider if variations in the bodily content of calcium bear any relation to different spasmodic conditions.

In infantile spasmophilia the total calcium in the blood is low, but it is because the ionic calcium is low that nervous symptoms occur. In other forms of tetany there may be no diminution of calcium in the blood serum, but the existence of an alkalosis, and in these cases the chlorine, not the calcium, is the important factor in treatment by calcium chloride. The place of calcium in the etiology and therapeutics of symptoms has then to be considered along with other factors, none more important than those associated with the endocrine system, particularly the parathyroids—and with vitamins, particularly vitamins A and D.

The significance of the parathyroids in the production of tetany in young children is still uncertain, but it is unusual for the parathyroid gland to be the primary cause of the trouble.

The conditions which produce rickets and spasmophilia are those in which there is a lack of the fat-soluble vitamins A and D. Deficiency of these vitamins causes lack of absorption of calcium, an abnormally low blood content of ionic calcium, and spasmophilia in infants. The blood content of ionic calcium can be raised either by increasing absorption, this being the fundamental method, by administration of vitamin (by irradiation, cod-liver oil, irradiated ergosterol); or by hypodermic administration of Collip's parathormone, altering and utilizing calcium reserves in the body, especially the calcium in the bones. The latter is obviously almost always an undesirable method of attempting to raise blood calcium in young children.

Apart from infantile spasmophilia there appears to be no definite relationship between blood calcium content and spasmodic conditions, or a positive facial irritability. Thus in fifty consecutive examinations of blood calcium, taken in seventeen cases of chorea under treatment, I have obtained reports varying from 7.33 mg. to 13.5 mg.

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per 100 c.cm., the severity of symptoms bearing no relation to the blood calcium content. In asthma and various neuroses the figure varied from 7.0 mg. to 12.3 mg.

Equally difficult to estimate is the value of treatment directed towards increasing blood calcium. It may be said, however, that where blood calcium is approximately normal in amount—that is, from 9 to 11 mg. per 100 c.cm.—no benefit is to be expected from any form of such treatment; when blood calcium is low, the all-important measure in therapeutics is administration of vitamin D (ostelin, for example). Granted that the child is under satisfactory conditions, dietetic and hygienic, the administration of calcium chloride or of parathormone may or may not cause improvement where there is low blood calcium; with the addition of vitamin D the prospect of improvement is great. Parathormone alone may cause a marked rise, but, as already noted, there are drawbacks to employing this treatment in childhood, and vitamin D is a safer and more powerful agent.

RELATION OF THE ENDOCRINE SYSTEM TO SPASM

The relation of the endocrine system to the autonomic nervous system is very close and complicated. According to Eppinger and Hess² two types of individual can be distinguished in whom the normal balance of the sympathetic and parasympathetic divisions is disturbed—namely, a group displaying undue irritability of the sympathetic (sympathicotonia), and another group with undue irritability of the parasympathetic (vagotonia). In this latter group are subjects liable to anaphylactic conditions—asthma and certain skin conditions—and diminution or removal of symptoms in these cases is to be obtained by stimulation of the sympathetic. If, then, allergic individuals are vagotonic, remedies having this effect are likely to be beneficial.

The ductless glands, which act in closest association with the autonomic nervous system, are the suprarenals, thyroid, and pituitary. While the function of the suprarenal in health is very uncertain, the effect of stimulation in times of stress through the outpouring of medullary secretion or adrenaline results in stimulation of the sympathetic and of thyroid activity; the converse is also true. Both diminish undue symptoms of vagotonia. For our present purpose it is important to note that suprarenal activity depresses the broncho-constrictors and that the characters of reaction to the therapeutic administration of adrenaline differ considerably in different individuals.

Thyroid instability is very marked in childhood. When thyroid activity is strained or excessive, all metabolic processes are increased in rate, and response to all forms of physical and mental stimulation is exaggerated. This may occur with the onset of puberty, infection, digestive or other metabolic disorder, or as a result of emotional strain; one outstanding characteristic of resultant excessive thyroid activity is great emotional excitement.

Even minor degrees of endocrine disorder, however, may cause mental disturbance—intellectual and temperamental. Cerebral organization is primarily stimulated by sensory impressions, and all of these are associated with some degree of emotional activity. "But our sensory reactions are more true to type, more constant in relation to the objects and impressions that excite them, than are our emotional reactions."

EMOTION AND INSTINCT

Emotion is not only a mental, but also a bodily process; every kind of emotion is normally accompanied by widespread physical changes, and excessive emotion may produce various forms of physical stress, disorder, or actual disease. Further, emotion not only has a sensory, but also a cognitive quality, from which chiefly arises

the possibility and the danger of the artificial production of emotion. Healthy emotion may be regarded "as a mode of experience which accompanies the working within us of instinctive impulses," and healthy emotion has as an essential feature an impulse to action. We have the outstanding example of fear with the suprarenal preparation for flight. Excessive emotional excitement may cause physical strain or disease, which again may increase susceptibility to emotional reaction, or, on the other hand, physical disorder may in the first place lead to hyperemotional conditions. In this connexion digestive disorder, whether as a primary condition or secondary to infection and fever, is especially noteworthy.

Allusion has already been made to the relationship between emotion and instinct. While the dominating parental instinct is the "mother of both Intellect and Morality," its unwise expression may ruin the child mentally, morally, and physically. The human young require prolonged tutelage, a guarded lengthy youth wherein slow development and organization may occur. During youth experience should be gained by free activity without full responsibility. If instincts are unduly curbed, if inhibitions are prematurely developed, trouble is inevitable. If a natural, healthy, slow, emotional development is gainsaid, thwarted reactions and activities disclose themselves in abnormal forms or degree of emotion, general disorder, and often in forms of neuromuscular disturbance—spasm.

We may say, therefore, that consideration of many spasmodic conditions in childhood involves study of general mental and physical conditions, and that the result of such study along the lines indicated by the spasm present is likely to give important indications for both general prophylactic and therapeutic measures.

ASTHMA

The asthmatic is vagotonic, and consequently sympathetic excitors are likely to prove useful in treatment. Following on the subcutaneous use and proved value of adrenaline in the acute condition came the reintroduction of ephedrine, efficacious by the mouth. Both these agents depress the broncho-constrictors, cause vaso-constriction in the bronchial mucous membrane, have a pressor action on the circulation, and raise blood sugar. Ephedrine is best given on an empty stomach; the effect takes some thirty minutes to occur, and lasts about five hours. It has been reported that some persons fail to respond to ephedrine, that in some tolerance is produced, while in others serious collapse may follow.

During recent years I have employed very largely in treatment peptone injections followed by ephedrine administration and as long supervision as possible. In the course of such treatment in approximately fifty cases certain points of interest have arisen, the cases being those in which gross disease of the chest was excluded by x rays, diet adjusted when opportunity permitted, and focal sepsis attended to. The series was restricted to hospital patients, and while in most of them protein cutaneous tests were made, in none were the results of practical value. In many instances multiple positive reaction occurred for which the most desirable treatment appeared to be general rather than specific. It is not true, however, that skin tests in early childhood are valueless, for in several other asthmatic patients, whose cases are not here included, dramatic benefit followed on treatment suggested by positive skin reactions.

A positive Chvostek's sign is not rare in asthmatic children, most of whom are negative to this test, which, whether positive or negative, is no sign of the blood calcium content. Many cases of asthma with F.I. apparently consistently negative improve greatly under treatment by peptone and ephedrine. Some, on the other

hand, show no marked improvement, but in none does ephedrine cause serious "collapse." One of the effects of adrenaline or ephedrine administration is a rise in the blood pressure, and it is a common experience in cases which respond to ephedrine to find that from half an hour after its administration the systolic pressure is distinctly raised for some hours. There are, of course, many difficulties and fallacies associated with accurate blood pressure estimations, but, granted the confidence and ease of the child, these are not insuperable so far as general approximations are concerned.¹⁰ In the cases of the type which I have mentioned the average response in a child of about 8 to 13 years of age to 1/2 grain of ephedrine is a rise in systolic blood pressure of rather less than 20 mm. of mercury, the diastolic pressure remaining stationary. In cases which do not respond clinically to treatment by ephedrine, and whose F.I. is persistently negative, it may be found that there is no blood pressure response to the drug, and that even when its administration is reinforced by thyroid there is still no response, or almost none.

Other children suffering from asthma have a variable F.I. reaction and respond indifferently to ephedrine—that is, they have an occasional attack of asthma while taking the drug. They have little or no blood pressure reaction, and they suffer no "collapse."

Lastly, there are some children who, a short time after taking ephedrine, suffer from "collapse," weakness, dizziness. I have met with 7 such cases—5 severe and 2 slight. In all the cases in which the collapse was severe, marked F.I. was present; in the mild cases it was absent. In all 7 cases the rise in systolic blood pressure was very marked after administration of ephedrine, the diastolic pressure remaining approximately level. In one case over several estimations the average rise was 26 mm. Hg, in another 30 mm. Hg, and in a third 40 mm. Hg. Symptoms can usually be avoided by smaller dosage. As a part of prophylactic treatment the prolonged administration of ephedrine is usually beneficial and always safe. It is most usefully given thrice daily before meals in a dosage of approximately 1/4 grain to a child aged 5 years and 1/2 grain to a child aged 10 years. When combined with careful and prolonged supervision—the most important element in treatment—the prognosis is good in early uncomplicated cases.

CHOREA

Chorea is a symptom, not a disease. When we consider the various conditions occurring at the different periods of life with which chorea may be associated, we find that all "have this in common, that the victim is not in possession of fully developed mental powers."¹¹

Chorea occurs in the young; it may result from the existence of intracranial lesions; or it may owe its origin to constitutional nervous instability. In all cases its onset is determined mainly by the emotional side of mentality. In childhood the chief importance of the symptom lies in the fact that it is usually associated with rheumatic infection. At this period of life some degree of nervous instability is natural. For the proper development of emotional control suitable experience is necessary. Lack of, or abnormality in, education and training may lead to emotional disorder, and may also form the basis of faulty motor control.

In the treatment of chorea many drugs have been tried, vaunted, and discarded. Notice is taken here of only two methods of treatment: the one directed to alteration of calcium metabolism and the other to nervous control by means of nirvanol.

My experience in regard to rheumatic chorea is that diminution in the amount of blood calcium cannot be an

important factor in determining the symptom. In fifty recent examinations the reports revealed that in twenty-one cases the blood calcium was between 9 mg. and 11 mg. per 100 c.cm.—that is to say, within the limits of the normal; in twenty-one instances it was below normal, and in eight above normal.

If, as was the case, very severe and acute chorea occurs with blood calcium figures above normal, the possibility that lack of blood calcium is an essential element in the development of the symptom is excluded. My evidence further goes to show that methods directed towards increasing ionic calcium have no marked influence on the symptom, and that they do not, in fact, in many cases increase the blood calcium content.

The methods used have been oral administration of calcium chloride and ostelin, and hypodermic injection of parathormone with ostelin by the mouth. On the latter treatment the blood calcium of a boy, aged 9 years, rose from 7.2 mg. to 11 mg. per 100 c.cm. in the course of three weeks, but the chorea remained very severe. In another case treated with parathormone, calcium chloride, and ostelin, the blood calcium figure during a period of six weeks was between 9 mg. and 9.3 mg. per 100 c.cm., and yet the child steadily improved.

A long-recognized method of treating choreic children has been by means of nerve sedatives. Of drugs recently introduced, nirvanol was found very useful in Germany for the treatment of various adult conditions, and it has later been largely employed for the treatment of chorea in childhood. It appears that in order to have any beneficial effect this drug, which is closely allied to luminal, has to be given in dosage sufficient to produce a somewhat violent reaction—"nirvanol sickness"—and that excessive dosage is dangerous.

In a series of ten consecutive cases of chorea treated with nirvanol, there was a good result in five. Although improvement was speedy and without distressing symptoms, it was accompanied by rash, pyrexia, and some initial exacerbation of symptoms. Persistence of improvement, however, was no more marked than with various other methods of treatment. In two of the other five cases great exaggeration of symptoms called for cessation of treatment. In one case hyperpyrexia with glandular enlargement was followed by rapid improvement; in the two others improvement was slow, and the ultimate results unsatisfactory. While nirvanol is sometimes associated with very satisfactory results, its use is severely restricted by the reaction it evokes, which is not entirely without danger.

FITS

The symptom of epilepsy occurs in various forms in childhood other than those of typical grand or petit mal, or of Jacksonian epilepsy. Early diagnosis is often difficult. A commencing idiopathic epilepsy may show itself for a long time solely by recurrent attacks of visceral spasm, and it may be only with the advent of an obvious fit, when the condition is fully established, that anti-epileptic treatment is begun.

Three common causes of fits due to gross intracranial abnormality are birth trauma, infantile cerebral paralysis, and amentia—for example, syphilitic meningitis. Epileptiform attacks may also occur unassociated with any gross intracranial abnormality as a symptom of, or in conjunction with, simple primary amentia, or they may be of the nature of idiopathic epilepsy. Since an ament is prone to fits, it is fundamental for prognosis to recognize whether amentia is present or not.

In a recent review of 129 children of school age noted as being sufferers from "epilepsy" I found that in 35 there was evidence of an organic cause or of primary

amentia; that in 16 out of the 94 cases of idiopathic epilepsy the patients were imbeciles, while 16 out of the 35 cases of organic lesion of the brain or primary amentia were imbecile. When a child has neither gross organic abnormality nor primary amentia but takes fits, these may have very different etiology and prognosis, and call for very different treatment. Epileptiform attacks forming part of the picture of spasmophilia—Group 3—are associated with rickets; they are intimately connected with the metabolism of calcium and phosphorus, are controlled by calcium injection, and can be prevented and cured by means of vitamin D. The ultimate prognosis here, for the mental condition also, is good.

Fits that may be classified under Group 4 are of certain types—for example, (a) infantile eclampsia, (b) an occasional fit or fits, (c) pyknolepsy, and (d) characteristic idiopathic epilepsy. In all, the presence or absence of an essential or idiopathic epilepsy may be conjectural for a time, but in none may the matter be lightly regarded. Although we have definite signs in spasmophilia for an independent diagnosis, and a definite exciting cause, the importance of a possible underlying diathetic trend towards convulsion is even here hard to evaluate.

When dealing with cases of infantile eclampsia we are on even less sure ground. Under this term I include the type of convulsions occurring during the first few weeks of life, usually in boys, where there is no apparent cause and often no digestive disorder, where the fits are of the nature of short general convulsions repeated at brief intervals, where other treatment is unavailing, but where temporary chloralization produces a permanent cure. Whether or not we have here to deal with an anaphylactic phenomenon is not known.

With a child who has an occasional fit we cannot altogether dismiss the possibility of an underlying vice of nature, provided there is no obvious cause, such, for example, as alkalosis. There is no certain test. But family and personal history, precedent or accompanying symptoms, the general physical and mental condition, may all go far to confirm a diagnosis in one or other direction.

In the rare type of case described as pyknolepsy we find that a previously healthy child of about 4 or 5 years of age suddenly begins to have fits of the nature of mild petit mal, which occur very regularly and usually very frequently from the onset. There may be as many as a hundred daily. As they commence, so they continue. No treatment has any beneficial effect; clinically striking is the fact that in spite of long-continued frequent attacks there is no mental impairment and no physical ill-health. After a variable number of months or years the attacks cease as suddenly as they began.

In striking contrast to such a course are most cases of ordinary idiopathic epilepsy; here the treatment consists essentially in attempts to restore nervous tone, and this demands attention to all bodily functions, particularly digestion and excretion. Chloralization is not successful, but the regular and long-continued administration of bromide and probably luminal is of the utmost importance. It is also necessary to establish intelligent control of instinct and emotions, to develop good habits, and to inculcate obedience, reticence, and modesty. Careful guidance of interest, occupation, and scholastic instruction is helpful. In some cases many of the peculiarities of the epileptic are due rather to his training, or rather spoiling, than to his weakness itself. The child is commonly not only a victim of a neuropathic heredity, but suffers also from a psychopathic home environment. Loose control is highly deleterious: the more average-normal his training the better. The satisfactory care of chronic nervous or spasmodic conditions, especially of

chorea, asthma, and epilepsy, demands the co-operation of parents, nurses, teachers, and doctors, and continuity of treatment for a long time.

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SUBPHRENIC ABSCESS

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A subphrenic, or subdiaphragmatic, abscess is a limited collection of pus just below the diaphragm. Frequently gas, either from a perforated viscus or as a product of gas-forming bacteria, is also present in the abscess cavity.

H. L. Barnard,¹ in his excellent paper upon this subject, analysed seventy-six consecutive cases, and classified subphrenic abscesses as follows:—

Intraperitoneal: right anterior, right posterior, left anterior, left posterior; of these, anterior abscesses are more common than posterior, especially on the left side.

Extraperitoneal: right, left; of these, the right-sided is much more frequent.

The falciform ligament is the dividing line between the right and left intraperitoneal abscesses, and the lateral ligaments of the liver separate the anterior from the posterior varieties. The right extraperitoneal space under the diaphragm is small, and is limited by the coronary ligament of the liver; the left extraperitoneal space is near the upper pole of the kidney. Extraperitoneal abscesses, however, may travel upwards behind the peritoneum from the colon, appendix, or kidney, pushing the peritoneum forward and eventually occupying the large space under the diaphragm. Sometimes a subphrenic abscess extends downwards into the subhepatic space on either side of the middle line. In the present series of 49 cases, 36 occurred on the right side, and 13 on the left.

CAUSES

Micro-organisms may travel by the blood stream from any part of the body and cause a subphrenic abscess, but the abdominal viscera are naturally the sources of most abscesses in this position, and of these the stomach and the appendix originate the large majority. The liver, gall-bladder, and bile ducts together contribute the next largest group of cases. Here the infection is usually spread direct from a perforated viscus, but it may be carried by the portal veins or by the lymphatics. Diseases of the chest, especially of the lungs, cause a few subphrenic abscesses; diseases and injuries of the chest wall, particularly of the ribs, are also etiological factors in a few cases. Either systemic or portal pyaemia may give rise to a subphrenic abscess. Many different organisms have been found in these abscesses, notably the *Staphylococcus aureus*, streptococcus, pneumococcus, *B. coli communis*, *B. pyocyaneus*, and *B. typhosus*. Subphrenic abscesses are most frequent between the ages of 20 and 50, and they are distinctly commoner in men than in women.