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ASSOCIATED MOVEMENTS OF THE HEAD AND EYES IN INFANTS.¹

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AND

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THE various forms of movements of the head and eyes in infants—the so-called rotary and nodding spasms of the head and vertical, horizontal, and rotary nystagmus—are observed quite commonly. The obscurity of their origin makes them especially interesting. For this reason Dr. Posey and I have decided to present these three cases, which happen to be attending our services in the Howard Hospital at the present time.

CASE I.—Carl C., aged three years, first came to the children's clinic in the Howard Hospital on October 13, 1903. The history recorded at that time was as follows: His birth was normal, and his first dentition complete. His parents are living and well, save that his mother is thought to be nervous. He has no brothers nor sisters, and his mother has had no miscarriages. His previous health has been excellent. He has had none of the infectious diseases of

¹ Read before the Philadelphia Pediatric Society, April 12, 1904.

childhood, and has never had any convulsions. For some time he has complained of abdominal pain. For many months he has lived chiefly on two cups of weak tea with each meal, milk, bread, and butter. He never eats meat. He eats freely of ice cream, but of other sweets rarely.

The condition which brought him to the dispensary was a cough of two days' standing which was accompanied by fever at night. The cough had a tendency to be paroxysmal, and the paroxysms were sometimes severe; it was tight, and there was no expectoration. He had some epigastric pain, and his bowels had been loose for several days. He had several stools each day, which were yellowish and watery, but contained neither blood nor mucus.

The physical examination showed a well-nourished child. His tongue was heavily coated, and its papillae red and prominent. His frontal and parietal eminences were prominent; there was moderate thickening of the epiphyses at the wrists and ankles. There was some costochondral thickening and some retraction of the base of the chest, more marked on the right side; there was moderate dorsolumbar lordosis and decided outward bowing of the tibiae. His abdomen was prominent; its walls flaccid, and there was marked separation of the bellies of the recti. His spleen was not palpable; but his liver was felt a finger's breadth above the transverse umbilical line, about three inches to the right of the umbilicus. There were a few scattered moist rales over each lung. He had a fairly loud, low-pitched, blowing systolic murmur, best heard in the aortic area and transmitted especially well to the top of the sternum and into the subclavians, less well into the carotids. A murmur of similar character was heard over the body of the heart, at the apex, in the axillary region, at the angle of the scapula, and in the interscapular space. At the apex the murmur was the same in character as that heard at the base, but in this area it seemed to occur later in systole; the second sound was heard distinctly over the upper chest.

On November 15th he had a mild follicular tonsillitis. On March 10, 1904, he was brought to Dr. Posey's clinic in the Howard Hospital on account of movements of the head and eyes and reduction in vision. These manifestations were first noted early in December. They had increased persistently up to this date. The head movements were lateral and not constant, the head apparently being drawn to the right and the chin slightly elevated, as if by action of the left sternocleidomastoid muscle. The eye movements were also from side to side and constant, the excursions being fairly extensive.

He was unable to recognize objects at a distance of four or five feet, but when brought within this range they could be defined. Any attempt at fixation caused a decided increase in the excursions of both the head and eye movements, especially the latter. Covering the eyes produced a temporary cessation of the movements of

the head, and they ceased during sleep. There was some restriction to the movements to the right in both eyes.

An examination of the eye-ground by Dr. Posey showed some engorgement of the veins, but no other abnormality. The only other nervous phenomenon noted was a slight uncertainty in his gait, and a mild ataxia, which had never been sufficiently marked to attract the father's attention. His reflexes were normal.

Further questioning showed that in December he had had a slight discharge from one ear, which cleared up quickly. About the same time he developed a cough, which has persisted. He has occasional severe paroxysms, which the father says sound like whooping-cough. Usually it is loose, but without expectoration. He vomits during or immediately after almost every meal. He does not seem to be nauseated, and frequently eats immediately after vomiting without being sickened. He never vomits at other hours of the day. His bowels continue loose—four or five stools each day—which are watery and yellowish in color. He voids urine frequently and in large quantity. Two examinations of the urine showed nothing abnormal, except a lowered specific gravity—1014 and 1012. Dr. F. R. Packard kindly examined the ears, throat, and nose, and reported nothing abnormal.

CASE II.—R. E., aged seven months, came to the Howard Hospital Dispensary on March 24, 1904. His birth was normal; his parents, three brothers, and one sister are living and well. One brother died aged two years of "baby consumption." He has had no convulsions, no manifestations of syphilis, and no previous illness. He has been fed exclusively on the breast about every third hour.

He was brought to the dispensary on account of a coryza of one week's duration; the discharge was free, and consisted of thin mucus without blood. His respirations were snoring in character—a condition which had existed since birth. There were no evidences of sore throat, but he had a cough in the beginning of the attack which was croupy in character. He has had no gastrointestinal symptoms.

The mother first noticed shaking movements of his head and eyes between the third and fourth months, and for some time he has had profuse head sweating. He is thought to have gained steadily in weight. He has always been a mouth-breather, and constantly chokes while nursing, as if there was some obstruction to nasal breathing. He seemed quite bright, and had no fever.

The physical examination showed a fairly well-nourished infant. There was no evidence of nasal membrane. His uvula was swollen and red. The fauces and tonsils were red, but without exudate. His tongue was moderately coated. His head was narrow in the bitemporal diameter, and the posterior fontanelle was still open. The coronal, sagittal, and lambdoidal sutures were wide open, and there was marked craniotabes along the borders of the squamo-

parietal and mastoparietal sutures, especially evident on the right side. He had a bald spot over the occiput. His neck was short, and the sternum elevated. His clavicles were on a direct plain with the body, and probably elevated slightly at the sternoclavicular junctions. His chest was markedly violin-shaped, his costochondral junctions were much enlarged, suggesting on palpation a backward dislocation of the cartilages. The portion of the chest wall between the costochondral junctions was pushed forward. The epiphyses were thickened at the wrist and knees, and there was slight dorsolumbar kyphosis. His abdomen was prominent, the bellies of the recti muscles were separated, and there was some pouting of the umbilicus. He had a frequently recurring spasm of the muscles of the back of the neck, with retraction of the head. Similar muscle spasms were not noted in other groups. His inguinal, axillary, epitrochlear, suboccipital, postcervical, and a few of his deep cervical glands were enlarged. He had marked lateral and tilting to-and-fro movements of the head, the head rotating to both sides as if by alternate contractions of the sternocleidomastoid muscles.

Dr. Posey's report upon the eye condition showed "lateral nystagmic movements with very short excursion and of similar extent in each eye. There were no apparent ocular palsies. On near fixation the movements were much increased, and this was associated with increased movements of the head. The examination of the eye-ground showed nothing abnormal."

On April 8th it was noted that the infant had emaciated decidedly, although his general condition seemed better; his head and eye movements were less.

CASE III.—Liliat B., colored, aged eleven months, applied for treatment at the Howard Hospital Children's Dispensary on August 13, 1902. Her birth was normal.

Family History. Both parents are living and well, but a paternal aunt and uncle died of phthisis. The child had had no previous disease, but had always been "poorly." She had been breast-fed for the first four weeks of life, since which time she had been fed on cows' milk, bread, crackers, cakes, candy, ice-cream, chocolate, and potatoes. She was brought to the dispensary for a cough, which she had had for three or four months. She was sometimes feverish, and had occasional night-sweats. Her cough was most severe at night. Her bowels moved six or seven times a day, the stools being yellowish in color, very loose, malodorous, and containing curds, but no mucus.

Physical Examination. The child was emaciated. The anterior fontanelle was very large, the sutures open, and the posterior fontanelle still open. The chest was markedly violin-shaped, and there was beading of the ribs. There was some epiphyseal thickening, the abdomen was very prominent, the umbilicus pouted, and there was separation of the bellies of the recti muscles. The

heart's action was regular and rapid, and the sounds clear. The breathing was labored, and the respiratory movements were increased in frequency. There were no areas of dullness. There was a wheezing sound on inspiration. The chest was full of sibilant and sonorous rales, by which the respiratory murmur was masked. There were no areas of bronchial breathing. The spleen was palpable just below the costal margin, and the liver one-half inch below. The cervical, anterior and posterior, the axillary and inguinal glands were moderately enlarged. The body temperature was normal.

The child made three return visits from this time up to the middle of October, 1902, and then disappeared until February 24, 1903. She was brought back on account of the return of the cough of the previous winter. She had recently had an otitis media on the left side. At the time she returned she was having a great deal of nasal discharge. Her appetite was poor, her cough loose, and attended by considerable expectoration. She slept well, but sweated a great deal about the head at night. It was noted at this time that she had a marked lateral nystagmus, which was said to have existed since the third month. It was much more marked in the right eye, and was at first thought to have been unilateral. There was no history of head movements at any time, nor of muscular twitchings in any other part of the body. At the age of sixteen months she had only her upper and lower middle incisor teeth. The anterior fontanelle was as large as a dollar, and the sutures were separated some distance from the fontanelle. There was marked bowing of the bones of the forearm, eversion of the legs, without much bowing of the bones, except a slight anterior bowing of the tibiae. The spleen was palpable a finger's breadth below the costal margin. The child was very weak, and required considerable assistance in order to maintain an erect posture. There were a few rales over the chest. The physical examination otherwise showed no changes.

Following this the child was absent from the dispensary for about eight months; when she returned with an aphthous stomatitis. Five months later she returned suffering from prolapse of the rectum. She was having three or four stools daily, accompanied by straining. They contained mucus, and there had been a slight amount of blood in some stools. This developed sometime after the prolapse of the rectum. Prior to this time the bowels had been normal. The abdomen was very much distended. There was no vomiting. The spleen was not palpable, the liver was two inches below the costal margin. There was no evidence of prolapse of the rectum found on examination.

The child returned twice following this, the last time on March 30th, on which occasion it was announced that there was no longer any prolapse of the bowel. A careful physical examination showed nothing to account for the prolapse. She had never vomited. She had no abdominal signs of any significance.

She is twenty-six months old at the present date. Her eye and stomach teeth are just erupting. All of her teeth are in poor condition and immaturely developed. One lower incisor has been lost by being caught in the thread of her skirt, indicating that the attachment of the teeth is insecure. The examination of the eye condition by Dr. Posey shows that when at rest the nystagmus is limited to the right eye; the movements are lateral, and the excursions very short. On fixation, or when the eyes are removed from the primary positions, slight nystagmic movements appear in the left eye also. There does not seem to be any limitation in the movements of the eyeball in any direction. Once or twice during examination, when the attention was suddenly attracted, there was a marked vertical vibrating spasm of the upper lids. Excitement increased the nystagmic movements, but the movements were not increased when the head was held fixed.

The question of the *etiology* of these phenomena remains unsettled. Numerous theories have been advanced, and the more recent writers have been in some accord in relation to this point. That the condition is dependent upon some functional disturbance of that part of the nervous system controlling the movements of the head and eyes is generally conceded. While it is not our purpose to treat this subject exhaustively it may be well to briefly review some of the more important theories that have been suggested.

Predisposing Causes. Age is an essential predisposing factor, the vast majority of cases occurring between the ages of two and twelve months, although Gee has reported a case occurring as early as the sixth week, and Henschel one as late as the third year. The cases are pretty evenly divided among the sexes with a slight majority in favor of females.

Heredity is supposed to play some part, in that a number of cases have occurred in families of bad nervous history, including epilepsy.

Rickets is probably the most common predisposing factor. In his early series of cases Hadden found this condition present in but two of eleven. On the basis of this finding he attached but little importance to it. In his second series of nine cases he found it present seven times, after which he modified his views decidedly. Other authorities differ as to its etiological relationship, the majority considering it an extremely common factor. The remarkable frequency of rickets, especially of its milder forms, would lead us to believe that its association has probably been closer than statistics would indicate. We are of the opinion that the milder manifestations of rickets, the head sweating, delayed dentition, and moderate heading of the ribs, but more especially the muscle and nervous phenomena, are of very frequent occurrence, and that, because we are prone to depend upon the skeletal deformities for our diagnosis, the manifestations in the nervous system—irritability, night terrors, etc.—and the muscle phenomena—pot-belly, gastric and intestinal

distention, constipation, etc.—are apt to pass unrecognized. In the dozen or more cases seen by one of us rickets of some degree was manifest in each.

Exciting Causes. *Dentition* was thought by Henoch to be the most common, because the conditions occurred during the period of first dentition, and because in some of his cases its beginning was coincident with the eruption of some of the first set of teeth. Most authorities admit that it may act as a contributing cause, but that it does not occupy the important position attributed to it by Henoch is manifest from the fact that many recorded cases have occurred at an age which would eliminate the teeth as a disturbing element.

Gastrointestinal irritation has preceded its occurrence in many cases, and it is generally conceded to be an important factor. It may act either reflexly or through the medium of toxic products absorbed from the intestinal tract.

Traumatism was recorded in a number of Hadden's cases, and in those of Peterson and Hirsch. Peterson thinks it probable that concussion is the determining factor in most cases—"a concussion setting up in the centres of the spinal accessory, and at times in those of the oculomotor nerves, an arrhythmic discharge."

Our knowledge of the pathology of these movements, for obvious reasons, is mainly speculative. Hugblings Jackson considered the condition a variety of "spinal system chorea, a symptomatic condition akin to canine chorea," while Gunn believed it was in some way associated with epilepsy, basing his view upon the quite common occurrence of temporary loss of consciousness in these cases, and the presence of epilepsy and other nervous conditions in the families in which they occur. Henoch previously suggested the same view, and Hadden, whose cases were the basis of Gunn's suggestions, gave this opinion some consideration.

Henoch, in describing his cases, suggested that the concomitant existence of these head-and-eye movements "seem to indicate that the root nuclei of the spinal accessory and of the upper spinal nerves supplying the affected muscles of the throat and neck stand in a very close relation to those of the ocular nerves (occulo-motorius)," a relationship which has since been definitely determined. Hadden believed that "the seizures pointed to instability of motor centres above the nuclei in the spinal cord and fourth ventricle, therefore attributing the disorder to a functional or other disturbance of the cerebral cortex. The child has acquired certain voluntary or purposive movements of the head and eyeballs, but these have not as yet become thoroughly organized and fixed in the psychomotor areas of the brain, hence a dissolution takes place because of the inability of the strained cortical centres to stand the work to which they have been too early subjected." This opinion, quoted by Mills, is one which he believes "throws the best light on an obscure subject."

Aldrich, in his very able discussion of this condition, voices the same view as to the nature of the condition. As we have stated, while we are by no means certain that the nature of these conditions is definitely determined, we incline to accept Hadden's explanation.

These three cases differ to some extent in the character of the head and eye movements. In Case III. the head movements are lacking entirely, the nystagmus is mainly in the right eye, and, according to the mother's statements, has existed since the third month. A point of special interest in this case is the occurrence of the vertical movements of the upper eyelid on an attempt at sudden fixation, a feature which has been noted but once before.

In Case II. the movements began at a very early period—between the third and fourth months—and involved both the head and eyes. They were equal on both sides. There was present in this case an intermittent spasmodic contraction of the muscles of the back of the neck.

In Case I. the movements began at an unusually late period—after the time that such movements have ordinarily disappeared. The head movements in this case are caused mainly by spasmodic contraction of the left sternocleidomastoid muscle, and the eye movements are the same on the two sides. In addition, there is some evident defect of vision and a slight ataxia, a somewhat similar condition having been noted in one of Hadden's cases. This is the only case in which it was possible for us to demonstrate any limitation in the excursions of the extraocular muscles.

Strabismus was not noted in any of the cases. There have been no antecedent nervous conditions from which the head and eye movements might have resulted, and none of them have had any of the acute diseases of childhood except Case I. The pupils were of normal size and reacted normally in all three cases, nor were there any changes in the fundi other than the dilatation of the retinal veins noted in Case I. The eyes in all three cases were moderately hypermetropic.

While the head-and-eye movements are manifested in a somewhat different way in each of these cases, and while we have been unable to elicit a history of head movements in one of them, we are, nevertheless, of the opinion that they are all dependent upon some form of irritation of the cortical areas.

The extreme rachitic manifestations present in all the cases marks this condition as the underlying or predisposing factor. The exciting cause in Cases II. and III. are difficult to determine. They both developed before the age of dentition, there is no history of injury, and they did not suffer from acute diseases which might have acted reflexly or by the development of some toxic irritant, or by suddenly producing increased instability of the motor centres, from the resulting exhaustion. The early age at which they became

manifest and the striking rachitic condition which was present lead us to believe that the toxic products resulting from the disturbed metabolism due to the rickets were the irritating factors in these cases. In Case I., however, the conditions are different. The rachitic manifestations in the child were much later, but very marked, quite sufficient to interfere with the nutrition of the nervous system.

The movements began in this case about two weeks after an attack of bronchitis, which was associated with diarrhœa, the latter condition persisting up to the present time. Furthermore, this child has been partaking most intemperately of tea for many months, regularly drinking two cups with each meal. The bronchitis, diarrhœa, and tea drinking, therefore, acting in a markedly rachitic child, we consider quite sufficient to have produced the phenomena noted.

A point of interest in these cases is the relationship which exists between the movements of the eyes and head. In an article which was read by one of us before the fifty-third annual meeting of the American Medical Association, entitled "Associated Movements of the Head and Eyes," the close association which exists physiologically between the movements of the head and eyes was dwelt upon, and it was shown how the head by its movements augments the range of the field of vision and supplements the action of the extraocular muscles in the delicate task of maintaining proper visual axes. Attention was also called to the fact that in order that the head may properly perform this twofold task it is provided with numerous and intricately acting muscles, and it is only when one considers that there is scarcely a position assumed by either the eyes or head in which the action of both parts does not enter that one realizes how intimate the relationship between the extraocular and cervical muscles is.

To facilitate the study, all associated movements which exist between the head and the eyes were divided into four groups. The first group referred to movements which are physiologic and will require no further mention here. The second group included abnormal simultaneous but independent movements, such as occur, for example, in disseminated sclerosis, where the movements which are abnormal and often simultaneous and have the same pathogenic origin are not related in the sense of being dependent upon one another. The third group dealt with compensatory movements. To this class were relegated cases which assumed a vicarious position of the head to compensate for some imperfect action of one or more of the extraocular muscles, such, for example, as occur after paralysis of one or more eye muscles. The fourth group referred to related but not compensatory movements, and included movements of the head and eyes which are abnormal and bear a distinct relationship to one another without being dependent upon

one another. Cases of head-nodding in children, such as have been presented to the Society to-night, were included under this latter group, although it was realized that there are a number of reasons why they should have been placed under the third group and the movements of the head considered to be secondary to those of the eyes. Thus, it is a well-established clinical fact that movements of the head, apparently similar to those observed in this class of cases, may originate in children whose eyes oscillate as a consequence of lowered vision, occasioned by a disease of the eyes, and it has been noted not infrequently that nystagmic adult patients hold their heads in a peculiar position while fixing for both distant and near objects, and often succeed in steadying the oscillation of the eyeballs in this manner. It has been remarked, too, in cases of the affection under discussion, by both Raudnitz and Caillé, that if the eyes are handaged the motion of the head ceases. Raudnitz noted also, in common with other observers, that the head movements were much influenced by the deviation of the visual axes, that they ceased when the eyes were deviated in certain directions, and started up again as soon as they were rotated from this position. This writer thought that the disease was particularly apt to arise in the dark winter months and to affect those confined in darkened rooms, believing that the head-jerking was secondary to the nystagmus, the latter being occasioned by strain upon the eyes consequent upon the infant's constant attempt to look toward the windows. Notwithstanding these observations and the well-established fact of the possibility of movements of the head being inaugurated by those of the eyes, it is probable that such is not the case in head-jerking, the movements occurring in association by reason of a common cause affecting the centres controlling each simultaneously, but without being dependent upon one another.

While in most instances the data which have been gathered regarding the time of the occurrence of the nystagmus and of the head-jerking are obtained by questioning the parents, and are, therefore, not entirely reliable, still there is evidence to show that the nystagmus does not always precede the head-nodding, that the head-nodding may even be observed some weeks or months before the jerking in the eye muscles manifests itself, and that, finally, head-jerking may occur without the occurrence of nystagmus during the entire course of the disease.

Moreover, in order that the movements of the head should compensate and neutralize those of the eyes, they should be directly opposite to one another; such a condition, however, has been found in but one case, the motion of the head being either in the same direction as those of the eyes, or, what is more usual, without any connection with them whatsoever. Again, the movements of the eyeballs are usually twice as rapid as those of the head and have their own rhythm.

It has been suggested that the presence of a strabismus in some of these cases would indicate that the nystagmus, under such conditions, at least, was the primary factor, but while the strabismus may indicate a palsy of one of the eye muscles nystagmus does not follow palsy of the eye muscles under other conditions.

A STUDY OF A CASE OF SPLENOMYELOGENOUS LEUKÆMIA.

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(From the medical service of Dr. Walnwright.)

THE subject of this study was previously under the care of Dr. J. Emmet O'Brien, of Scranton, and Dr. Thompson, of Carbondale, to both of whom we are indebted for notes of his former history.

The patient stated that about six years before admission to the hospital he had a severe attack of abdominal pain, with vomiting and diarrhœa. He said that this illness was similar to his subsequent ones. He was given arsenic at the time, and made a good recovery, and was apparently in good health until late in the year 1902, except for a time in 1901, when he was bothered by a pain in the splenic area.

About October, 1902, he began to suffer again with "bloating" and with alternate constipation and diarrhœa. Dr. O'Brien first saw him on December 11, 1902. He found the spleen much enlarged. A record of the blood count was not kept, but it is known that the leukocytes were much increased in number, and smears showed the picture of a leukæmia of the splenomyelogenous type. Fowler's solution in increasing doses was prescribed. On January 3d the patient returned much improved, and the splenic tumor was not palpable. On January 28th the spleen could again be felt, and the leukocytes were still numerous. His general condition, however, seemed to be improved. He was last seen by Dr. O'Brien on March 24, 1903.

From March, 1902, until his admission to the hospital he was seen a few times by Dr. Thompson, who was called chiefly for gastrointestinal trouble. One of these attacks yielded to small doses of calomel daily. At another time he was again given arsenic, with benefit.

On the whole, during this period his condition was fairly good. He seems to have been troubled only by the attacks of diarrhœa, and he kept very steadily at his active duties as inside foreman of a coal mine until March 31, 1904.