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CONGENITAL FACIAL PARALYSIS.*

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For the last few years a great deal of interest has been taken in congenital abnormalities of muscles. The absence of one or more muscles, or parts of muscles, has been noted from time to time, and although such a congenital absence is by no means common, still a considerable number of cases has been reported.

The pectoral muscles seem to be particularly prone to such a defect in development, as by far the greatest number of reported cases has to do with these muscles. Hoffman,¹ from Eichorst's clinic, and Kalisher² have collected and analyzed these cases. Many of the cases showed other congenital abnormalities, such as malformations of the skin and hair, webbed fingers, etc., etc.

Möbius,³ in 1888, and again in 1892, called attention

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¹ Hoffman: Virchow's Archiv., vol. cxlvi., 1896, p. 163.

² Kalisher: Neurolog. Centralbl., vol. xv., 1896, pp. 685, 732.

³ Möbius: München. med. Wochenschr., xxxv., No. 6, 7, 1888; ibid., xxxix., No. 2, 4, 1892.

to an interesting group of cases, in which there was a congenital defect in the movements of the eyes, combined, at times, with a similar defect in the facial muscles. With these he associated certain cases which developed in infancy or childhood, and called them all infantile nuclear atrophy (*infantilen Kernschwund*), for he believed that the process was essentially the same in all the cases—a degeneration and atrophy of the nuclei of the cerebral nerves.

The muscles of the eye were affected in all of the cases that Möbius collected. That the facial muscles might be affected alone, he thought was probable, although he had been unable to find any record of such cases.

Schultze,⁴ in 1892, and Bernhardt,⁵ in 1894, report cases of unilateral congenital facial paralysis. I shall refer to these cases again.

Kunn,⁶ of Vienna, in 1895, in a monograph considers the congenital defects in the movements of the eyes. (*Die angeborenen Beweglichkeitsdefecte der Augen*.) He has abstracted and tabulated all the cases he could find in literature. Among these 73 cases, there were 11 in which the facial muscles were also involved. He refers to several cases of uncomplicated congenital facial muscular defect. As opposed to Möbius, he makes a sharp distinction between congenital cases and those which develop after birth, and, as the anatomical basis for the congenital cases, he assumes a lack of development somewhere in the motor path from the brain to the muscles.

Last year Schmidt⁷ reported a case in which there was an absence of the left pectoral major, combined with defects in the muscles supplied by the XII., VII. and VI. cerebral nerves.

⁴ Schultze: *Neurolog. Centralbl.*, xi., 1892, p. 425.

⁵ Bernhardt: *Ibid.*, xiii., 1894, p. 2.

⁶ Kunn: *Beiträge zur Augenheilkunde*, Heft xix., 1895, p. 1.

⁷ Schmidt: *Deutsche Zeitschrift. f. Nervenheilk.*, x., 1897, p. 460.

From this short glance at the literature, it may be stated that congenital abnormalities of the muscles occur not very infrequently; that they are most common in the pectoral muscles and the muscles of the eyes, and that the facial muscles are at times implicated, usually in association with those of the eyes, but at times alone.

The facial defect, when it has been combined with that of other muscles, has been bilateral; when it has been uncomplicated, it has been unilateral.

As far as I have been able to discover, a satisfactory case of congenital facial diplegia has not, or, rather, had not been reported until I showed one of my cases to the Johns Hopkins Hospital Medical Society. The two cases which I have observed have an added interest, as they are brothers.

Seth O., æt. 21; Basher O., æt. 19.

The parents of these two boys are strong, healthy country people. There is no history of alcoholism and no reason to suspect syphilis. The mother's aunt gave birth to a child with only one arm, and the mother herself had a baby with a deformed foot. This child lived only two hours. Other than this there is no history in the family of congenital deformities. The mother accounts for the condition of her boys by "maternal impression", as she had been very much affected, while pregnant with the first boy, by hearing of a minister in her church who was unable to move any of the muscles of his face. Before the birth of the second case she was very anxious for fear that this child would be born also with a deformed face. It may be remarked in passing, that her anxiety about the faces of the subsequent children, of whom there were three, all boys, had no effect upon the development of their facial muscles. An elder daughter is also perfectly normal. Three other children died in infancy.

Case I.—Seth O., æt. 21. The patient, a second child, was born without instruments, although the labor was difficult and protracted. His mother noticed soon after his birth, that his under lip drooped, and that he did not close his eyes when sleeping. He was able to suck without difficulty by the use of his tongue. When he was old enough to smile, his face remained expressionless. The patient learned to walk at thirteen months, and to talk when about two years old, although he has never learned to pronounce certain words distinctly.

Dentition began very early, the first tooth being cut when he was little more than six weeks old. He developed normally, learned to run and play games as other boys, and except for his expressionless face there was nothing to be noticed about him. He left school when he was twelve, as he then became sensitive as to his appearance. As a child he was ill with measles, and his mother thinks that his deafness is a re-



FIG. 1. (Case 1.)

sult of this disease, although he had no discharge from his ears, or any symptoms referable to it.

The examination (Nov. 3d, 1896) at his home in a neighboring State, shows him to be in general a well-developed young man, rather shorter than the average (Fig. 1.) His intelligence seems to be good, his speech is somewhat in-

distinct, as the labials cannot be given their proper sounds. B is pronounced D, F ech, M is N, V is ch or ge, the sound of W is poorly given. His face is perfectly expressionless, forehead smooth, without wrinkles, eyes wide open, mouth open, lower lip large and everted. The lobe of the left ear is misformed, there being a distinct division between the part next to the cheek and the rest of the ear. On the right side there is some indication of this abnormality. His teeth are

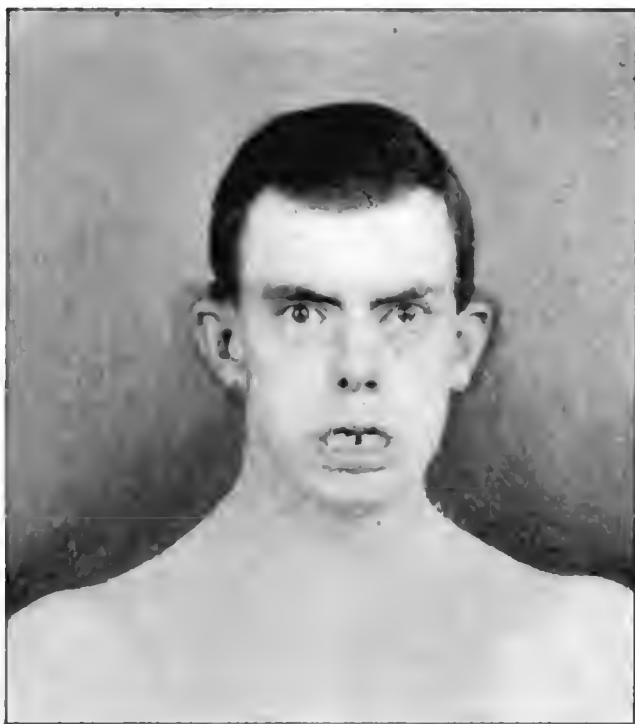


FIG. II. (Case 2.) Face in repose.

fairly well formed. The senses of smell and sight show no marked abnormality. His eyeballs are not especially prominent, are straight and freely moveable in all directions, and there is no nystagmus. The pupils are equal and react to light and during accommodation.

The muscles of mastication act well and equally on the two sides. Sensation in the face is normal, and the sense of taste is unaffected.

The patient is entirely unable to raise or to contract his eyebrows. When trying to close his eyes, the eyeballs are rolled up and the upper lids are somewhat relaxed. He is unable to elevate his upper lips or to pucker his mouth, or indeed to close it. On the right side he is able to draw the angle of his mouth somewhat outwards. This motion is impossible on the left side. He can depress and retract the angles



FIG. III. (Case 2.) Face in repose.

The muscles of the soft palate act normally; the pharyngeal reflex is active. He protrudes his tongue in the middle line, and is able to move it freely in all directions. The action of his mouth by the action of the platysmæ. An electrical examination could not be made. The patient is quite deaf in both ears, being just able to hear a loud-ticking watch upon contact.

his heart is normal in all respects. The muscles of his shoulder girdle, arms, hands, trunk and legs are well developed and of normal strength. The deep reflexes are normal, and no other abnormality of any kind is discovered.

Case II — Bashir O., *et. 10.*

Birth normal. The defect in the baby's face was noticed shortly after his birth. He was able to nurse, as was his



FIG. IV. (Case 2.) Showing extent of voluntary control of muscles.

brother, by the use of his tongue. He developed very much as his brother did. When he learned to speak, there was the same difficulty in pronouncing words, etc. He cut his teeth at the usual time. He was an active boy, fond of all out-door games. The facial defect never changed from his earliest infancy. He has learned to smoke, holding the cigarette in

his tongue, and, indeed, he has taught his tongue to do many of the services that are usually performed by the lips.

Present condition. Nov., 1896. Figs. II. to V.

The patient is a well-developed youth. His intelligence is good. His speech shows the defect noted in the case of his brother, that is, he is unable to pronounce the labials. His face is remarkably expressionless, eyes are wide open and



FIG. V. (Case 2.) Faradic stimulation of platysma.

prominent, mouth is held open, lower lip is large and pendulous. The lower jaw is protruded, the lower teeth being more than half an inch beyond the upper teeth. His teeth are poorly developed. The lobe of the right ear is notched, in a similar manner to that of his brother.

The patient's vision is normal, his eyes are in the median

line, freely moveable in all directions, and there is no nystagmus. The pupils are equal and react to light and during accommodation. The muscles of mastication are unaffected the sensation of the face is normal. Taste is acute on the fore part of the tongue. He is unable to elevate or contract his forehead in the least. When endeavoring to close his eyes, the eyeballs are rolled upward and the lower lids are relaxed (Fig. IV.). He cannot elevate his upper lip, but is able to retract and depress the corners of his mouth by the use of the platysma. The buccinators have also retained some power. Electrical stimulation from the root of the facial nerve causes contraction in the muscles which can be voluntarily moved, and in those moving the ear. By direct stimulation the platysma can be brought into play (Fig. V.). It takes relatively strong current to produce these contractions. The patient is deaf in both ears, and can hear a loud-ticking watch only when it is within two inches of his ears. The movements of the soft palate and tongue are normal. The pharyngeal reflexes are normal. The development in the arms, trunk and legs is excellent. Nothing else abnormal is noticed.

The patient entered the hospital in Dr. Halsted's wards and underwent two plastic operations devised to bring his lips closer together. In the first operation a bit of lower jaw was excised, and in the second the redundant portion of the lip was cut away. There was marked improvement, his lips being nearly approximated.

During the second operation it was noticed that there was very little muscular tissue in the lip, it being composed largely of fat. Fibres of what was supposed to be the platysma were made out.

That the muscle defect in these cases is a congenital one, I think cannot be doubted. In the first place, the mother is quite sure that the defect was present when the children were born, although she cannot state that she noticed the deformity immediately after birth. The fact that the malformation occurred in two members of the same family, and that it is bilateral, speaks for its congenital origin, and the occurrence of other faults in development, the misshaped ears, and, possibly, the deafness, lends added weight to this view. The character of the paralysis itself is quite similar to that which was found in the other congenital cases.

There are 14 cases of facial paralysis combined with

eye-muscle defect; these have been reported by Graefe,⁸ 1880; Harlan,⁹ 1881; Chisolm,¹⁰ 1882; Armaignac,¹¹ 1886; Möbius,¹² 1888; Schapringer,¹³ 1889; Bernhardt,¹⁴ 1890; Bloch,¹⁵ 1891; Fryer,¹⁶ 1892; Bach,¹⁷ 1893; Remak,¹⁸ 1894; Gesèpy,¹⁹ 1894; Schmidt (loc. cit.), and Procopovici,²⁰ 1896.

I have not included Rechin's two cases, because in the one the onset is said to have been in the fourth year, and in the other there was no true facial paralysis, nor Hanke's case, as the history of the onset is not satisfactory.

In these 14 cases, the defect in the VII. nerve was combined with that in the VI. no less than 12 times, and in 7 cases there were no other cranial nerves involved. In one, the sensory portion of the V. was also involved, and in another there was a partial defect of the motor V. and of the XII., and in 2 cases the external ophthalmoplegia was practically complete, involving the III. and IV. nerves as well as the VI. The fifth case is the interesting one reported by Schmidt, in which, combined with the pa-

⁸ Graefe: Case 4, Handbuch d. ges. Augenheilk., 1880, vol. vi., p. 60, cit. from Kunn.

⁹ Harlan: Case 1, Trans. of the Am. Ophthal. Soc., 1881, p. 216, "Congenital Paralysis of Both Abducens and Both Facial Nerves."

¹⁰ Chisolm: Archives of Ophthalmology, vol. xi., 1882, p. 323.

¹¹ Armaignac: Case 3, Rev. Clin. d'Oculistique, November, 1886, cit. from Kunn.

¹² Möbius: Münch. med. Wochenschr., 1888, No. 667, "Ueber angeborene doppelseitige Abducens Facialislähmung."

¹³ Schapringer: New York med. Monatschrift, December, 1889; Boston Med. and Surg. Jour., 1889, p. 635.

¹⁴ Bernhardt: Neurolog. Centralbl., 1890, vol. ix., p. 419, "Ueber angeborene einseitige Trigemini Abducens Facialislähmung."

¹⁵ Bloch: Berlin Thesis, 1891.

¹⁶ Fryer: Ann. Ophthal. u. Otolog., Kansas City, 1892, "Case of Congenital, Bilateral, Ext. Ophthal. and Cong. Bilateral Facial Paralysis," cit. from Kunn.

¹⁷ Bach: Centralbl. f. Nervenheilk., xvi., 1893, p. 57.

¹⁸ Remak: Neurolog. Centralbl., 1894, No. 7, "Ein Fall von einseitigem angeborenem Defect des Platysma myoides."

¹⁹ Gesèpy: Arch. d'Ophthal., xiv., 1894, No. 5, 273, "Deux Cas d'Ophthalmoplegie Congen. Externe."

²⁰ Procopovici: Arch. f. Augenheilk., xxxiv., p. 34, "Ueber angeborene, beiderseitige Abducens Facialislähmung."

ralysis of the VII. and VI. nerves, there was an unequal paralysis of the XII. pair, the left more than the right, and an absence of the pectoralis major muscle on the left side.

The defect was bilateral in all of these 12 cases, except in Bernhardt's case, in which the paralysis was confined to the right VII., VI. and V. nerves. Bernhardt believed that the condition was caused by some injury to the nerves at the base of the brain, which they had received during birth, and it is doubtful whether the case should be included with the others. I shall have to return to this case, in speaking of the pathology of the condition. .

The two cases in which the VI. nerve was normal are of particular interest. The first is that of Armaignac, in which there was a left-sided defect of the orbicularis palp., and probably also of the frontalis, combined with paralysis of the levator palp. and the superior rectus on the same side. Remak's case is the second. In this case the left platysma, including the quadratus and triangularis menti, was absent or paralyzed, and there was also a bilateral defect in the levator and the superior rectus.

As a rule, all the muscles supplied by the VII. were not affected. In Schmidt's case there was complete paralysis. The notes in Fryer's, Bernhardt's, Schaplinger's and Bach's cases are not specific on this point. Therefore, in the 10 cases in which there was a definite note, the paralysis was incomplete in 9. The muscles which draw the mouth outwards and downwards (the platysma, etc.) are particularly likely to be spared. They were spared alone four times; once the orbicularis palp. was also not affected, and in two other cases the paralysis was confined to the upper branch of the VII. In Bloch's case this paralysis was of the lower branch of the VII. In Remak's patient the platysma, the quadratus menti, and the triangularis menti were the only muscles paralyzed, involving just the muscles which are usually spared.

There are 6 cases in which there was an uncomplicated congenital facial paralysis. These have been reported by Stephan,²¹ Henoch,²² Schultze,²³ Bernhardt,²⁴ Procopovici²⁵ and Bernhardt.²⁶ I have not included Delprat's case (cit. after Möbius), as there is the definite history of the onset at three, after an acute illness; nor Kunn's case, as there is here a very different condition, a right hemiatrophy of all the structures of the face below the eye; bones as well as muscles.

In all of these, except in Procopovici's case, the defects were unilateral, three times on the left and twice on the right side. I shall speak of Procopovici's case a little further on, as it belongs more to the cases which I have been reporting than to these.

In all but Stephan's and Henoch's cases the paralysis was incomplete, and it was the muscles about the mouth that were spared. In fact, the clinical picture differs in no way, except in being unilateral and uncomplicated, from that of the VII. nerve paralysis in the combined cases. Bernhardt, in trying to answer the question as to whether these cases should be considered as strictly congenital, shows that they differ in no way from those which are caused by injury to the VII. nerve. That all the muscles supplied by the VII. nerve are not paralyzed, and that there is no secondary contraction, does not, as Kunn thought, serve to distinguish them.

Facial paralysis due to the injury of the nerve at birth is not a very uncommon accident; it is usually caused by the application of forceps, but may occur even during

²¹ Stephan: *Rev. de Med.*, July, 1888, p. 548, and *Nederl. Tijdschrift*, 1888, p. 113, cit. from Bernhardt.

²² Henoch: *Vorlesungen über Kinderh.*, 1897, ix. Auflage. Also abstr. Bernhardt, *Neurolog. Centralbl.*, ix., p. 423.

²³ Schultze: *Neurolog. Centralbl.*, xi., 1892, No. 14, p. 425.

²⁴ Bernhardt: *Neurolog. Centralbl.*, 1894, xiii., p. 2.

²⁵ Procopovici: *Arch. f. Augenheilk.*, 1896, xxxiv., p. 44.

²⁶ Bernhardt: *Neurolog. Centralbl.*, xvi., 1897, p. 296.

normal labor. ²⁷ ²⁸ This paralysis is, in the great majority of cases, a transient one, and the recovery is quite complete. In some cases, however, the paralysis does not get well, and the condition persists throughout life. When this is the case, the symptoms are identical with those described in the five cases under discussion.

As an example of this, I may give briefly the history of a case that I have had under observation for some time. The patient is a young man of excellent health, who, except for his facial paralysis, shows no abnormalities. He has been told by his parents that the condition was noticed directly after birth, and that his birth was very protracted and difficult. He thinks forceps were used, although of this he is not certain. He is certain, however, that the paralysis has always been ascribed to injuries received to his face during birth, and that the condition has not changed since he can remember. At present the left side of his face is almost completely paralyzed. The forehead cannot be raised nor the eye closed, nor the upper lip elevated, and the lips cannot be puckered. He can, however, draw the left angle of his mouth outwards, throwing the cheek into longitudinal folds. He is not able to contract voluntarily the platysma on either side. Electrical stimulation of the left VII. nerve causes contractions of the muscles, moving the angle of the mouth outwards, and nothing else. Another patient, a woman, 34 years old, who has a left-sided facial paralysis, gives the history of having had it since her birth, which was non-instrumental; but, as I have been unable to confirm the history, I shall simply mention it in passing.

Bernhardt, with great fairness, concludes that, although the occurrence of an isolated, unilateral, congenital facial paralysis, or, perhaps, better, an incomplete development of the nerves and muscles in the distribution

²⁷ Geyl: *Centralbl. f. Gynäkologie*, xx., 1896, p. 634.

²⁸ Knapp: *Ibid.*, xx., 1896, p. 705.

of the facial nerve on one side, cannot be denied, still, its occurrence has, as yet, not been definitely demonstrated.

Procopovici (loc. cit. p. 45) refers briefly to an interesting case, which he says had lately come under observation. It is that of a man, 18 years old, in whom there had been, since his birth, a paralysis of the upper branch of the facial nerve. All the muscles which are supplied by the facial nerve were active, except only the orbicularis oculi and the frontalis, which were paralyzed on both sides in almost equal intensity. In other respects the patient was well. (This is the full note.)

Procopovici refers also to another case in the footnote on page 44. A woman, aged 34, had been born with a paralysis, which was more intense on the right side. It affected the muscles of the forehead, the orbicularis palpebrarum and all the muscles of the face. The muscles of the soft palate were not affected. Meynert thought, on account of the distribution, that it was a peripheral paralysis of all the branches of the facial nerve, external to the Fallopian canal. Accidents during birth were to be excluded. The abducens was normal, and hearing was unaffected.

These are the only two cases that I have been able to find of uncomplicated bilateral congenital facial paralysis, and in these cases the histories are so meagre that it is difficult, or impossible, to definitely determine their character. The first of these cases appears to me to be particularly interesting, as the paralysis involves the upper branch of the facial nerve, the branch which is believed by some to arise near the nucleus of the third nerve. In this connection I shall recall Armaignac's case, in which there was a congenital defect of the orbicularis palpebrarum and of the frontalis of the left side, combined with a similar defect in the levator palpebrae and rectus superioris of the same side. In Gesèpy's case, in which there was a bilateral paralysis of the orbicularis palpebrarum, there was complete external ophthalmoplegia.

As for the pathological basis which underlies this interesting condition, very little can be definitely said. Its pathology is almost entirely speculative. Möbius, who was the first to make any exhaustive study of these cases, associated the cases of congenital paralysis of the facial and abducens nerves with the other cases of congenital paralysis of the eye muscles, and he brought these congenital cases into relation with cases which developed in infancy and childhood, and presented symptoms which were quite similar. That the congenital cases might be due to some defect in development, an aplasia of the motor apparatus, Möbius recognized, but he thought it was better to assume the same process for both the congenital and acquired cases, and this process he believed to be an atrophy of the nuclei from which the nerves arise.

Kunn, in his monograph, written several years after Möbius' second paper, reviews the whole literature, and makes a sharp distinction between the congenital and the acquired cases. He bases this distinction upon what he considers definite clinical differences; these are in relation to the paralysis of the ocular muscles, and do not particularly concern us at this time. He believes that the congenital cases are not due to an atrophy of the nuclei, but should be considered as a defect in the development in the motor mechanism, and he announces the theory that the defect may be anywhere in the motor path, from the cortex of the brain to the muscles. Kunn admits that this theory is based on a very slight anatomical foundation.

In congenital defects of the eye muscles, which have been operated upon, the muscles have at times been found wanting, and at times in every degree of development, up to what appeared perfectly normal muscles. In certain cases microscopic examination of a bit of excised muscle showed a condition quite similar to that which is described in progressive muscular dystrophy. In regard to the central nervous system, there are really no ex-

aminations that speak definitely as to the condition of the nuclei in these cases. Bernhardt examined the brain in a case in which the right VII. nerve, the VI. nerve and the sensory portion of the V., on the same side, were paralyzed. The defect was noticed shortly after birth. The child died when it was nine months old. Two foci of softening were found—one superficial in the right side of the pons, the other more extensive in the right corpora quadrigemina. The nuclei of the cerebral nerves were said to be normal. The peripheral nerves were not examined. Bernhardt himself believes that there was an injury to these nerves at the base of the brain during birth, and it is very doubtful whether this case should be included with the strictly congenital cases. Siemerling²⁹ describes the autopsy on a man who had had a congenital ptosis of the left eye, and who died from general paresis. He found a degeneration in certain cells of the nucleus of the third nerve. The degeneration was bilateral, and suggested rather a later process than one which had been in existence for fifty years. The character of the process, and the fact that the patient had general paresis, and that the lesion was bilateral, whereas the muscle defect was unilateral, would seem to justify Kunn's objection to considering this a conclusive case.

But it would seem to me that Kunn was probably right in distinguishing the congenital cases from the acquired cases, and, from the point of view of congenital facial paralysis, it is interesting to note that Möbius was unable to find a case of abducens-facial paralysis which developed in childhood. The case which he referred to as a doubtful case, that of John Thompson,³⁰ seems more likely to have been due to a neoplasm of the medulla than to a nuclear atrophy.

That congenital facial paralysis is almost always as-

²⁹Siemerling: *Arch. f. Psych.*, xxiii., 1892, p. 764.

³⁰Edinburgh *Med. Jour.*, vol. xxxvii., 1891, p. 262.

sociated with paralysis of the VI. nerve would lead one to believe that the mal-development is in the medulla, near the origin of these two nerves. That the nuclei of both nerves need not always be affected together is shown by the occurrence of uncomplicated congenital VI. nerve paralysis, and there seems no reason for assuming that such a condition could not occur in the nucleus of the VII. nerve, and that is what I assume has happened in the boys whose cases I have reported. If, in fact, a mal-development, or perhaps a non-development, of the nuclei accounts for the occurrence of these cases, it does not follow that the muscles themselves must be absent. The cases which Frl. v. Leonowa³¹ reported demonstrate that muscles may develop independently of the central nervous system. She examined two monsters which were entirely without brain or spinal cord. The dorsal root-ganglia and nerves growing from them had developed, as had the muscles. In many cases of congenital ptosis curious associated movements occur in relation to movements of the jaw, and this would also indicate that the defect was in the nucleus, and that this associated movement had its representation apart from the nucleus of the third nerve, probably in the nucleus of the V. nerve.

That there may be a congenital absence of the muscles, the cases of the absence of the pectoral and other muscles seem to prove, but it is hard to understand why such a defect should be strictly limited, as it was in our cases, to the distribution of one nerve, if the nervous mechanism had nothing to do in determining it, and when we associate with this, as so often happens, a defect in just that one of the six external muscles of the eye whose nucleus lies in close relation to the VII. nucleus, any other explanation than a defect of these nuclei seems very far-fetched.

Schmidt's case, in which there was a defect in the VI.

³¹ v. Leonowa: *Neurolog. Centralbl.*, 1893, pp. 218 and 262; 1894, p. 729.

and VII. nerves, combined with absence of the left pectoralis major muscle, does not demonstrate that the process was the same in the muscles of the face and the muscle of the chest. Congenital abnormalities are rarely unassociated with other malformations, and even if we assume a different cause for the absence of the pectoralis major and paralysis of the facial muscles, it would not be surprising that they both might occur in the same individual. We do not, however, know the cause of the congenital absence of individual muscles, and it may depend upon some fault in the development of the central nervous system.

There is no proof that a congenital abnormality in the upper motor segment could produce defects of this character, and I agree with Möbius that such a lesion is not to be assumed. In conclusion, we must acknowledge that we do not know the anatomical basis for the cases of congenital facial paralysis, but it is more in accordance with the known facts to assume some fault in the development of the nucleus of the VII. nerve to account for these cases.

FACIAL PARALYSIS COMBINED WITH EYE-MUSCLE DEFECT.

Graefe: Handb. d. ges. Augenheilk., 1880, vol. vi., p. 60.

Cit. from Möbius. Left VII. nerve paralyzed. Right upper and middle branches weak. Bilateral VI. nerve paralyzed. Smell and taste somewhat affected.

Harlan: Trans. of the American Ophthal. Soc., 1881, p. 216. Bilateral paralysis of VII. and VI. nerves (complete?). Taste normal. The platysma active. Slight downward movement of mouth.

Armaignac: *Révue clin. d'Oculistique*. November, 1886. Cit. from Kunn. Left paralysis of orbicularis palp. (frontalis probably), and levator palpebræ and rectus superioris.

Chisolm: *Archives of Ophthalmology*, vol. xi., 1882, p. 323. Bilateral paralysis of VII. and VI. nerves. Muscles about lower lip retaining some power.

- Möbius: Münch. med. Wochenschr., 1888, p. 667. Bilateral paralysis of VII. and VI. nerves. Some retention of muscles about mouth. Left side moved a little out while talking. Elect. m. m. which draw mouth out and down L. and R. are excitable. Also buccinators. Smell, hearing and taste normal.
- Schrapinger: Boston Med. and Surgical Journal, 1889, p. 635. Bilateral paralysis of VII. and VI. nerves. Paralysis of motor V. and XII. nerves. VII. nerve paralysis not specified as to extent. Other congenital abnormalities.
- Bernhardt: Neurolog. Centralbl., 1890, vol. ix., p. 419. Paralysis of right VII. and VI. nerves, also sensory disturbance. Infant, autopsy.
- Rechin: Klin. Monatsbl. für Augenheilk., 1891, p. 340. Cit. from Kunn. Two cases, doubtful. First said to have developed at four years. Bilateral paralysis of VII., especially lower branch. Nearly complete ophthal. external. Second case, facial muscles thin, badly developed, but not paralyzed. React normally to electricity. Ophthalmoplegia ext. All muscles of body badly developed.
- Bloch: Berlin Thesis, 1891. Case 28. Boy nine months old. Bilateral paralysis of VI. nerve. Bilat. paralysis of VII. nerve, lower branch. Bilat. club-foot and other abnormalities. The affection was congenital. (Note short.)
- Fryer: Ann. Ophthal. u. Otolog., Kansas City, 1892. Cit. from Kunn. Bilateral paralysis of VII. and VI. nerves.
- Bach: Centralbl. f. Nervenheilk., xvi., p. 57, 1893. Man 27. Bilateral ptosis. Bilat. ophthal. ext. Condition congenital. Facial muscles were flaccid. Frontalis and corrugator contracted. Eyes can be closed. Features expressionless. Lips moved only slightly during speech; no absolute paralysis. Operation on ptosis. Muscle normally placed. No noticeable abnormality.
- Rémak: Neurolog. Centralbl., 1894., No. 7. Paralysis of

- left VII. nerve, and bilateral paralysis of levator and superior rect. The platysma, the quadratus and triangularis menti were the only facial muscles affected. Gasèpy: Arch. d'Ophthal., xiv., 1894, No. 5, p. 273. Cit. from Kunn. Bilateral paralysis of VII. nerve (orbic. palp.) and ophthal. ext. nearly complete.
- Hanke: Wiener klin. Wochenschr., 1894, 46. Abstr. Schmidt's Jahrbücher, vol. 246, p. 22. Doubtful case. History of onset unsatisfactory. The "staring look" had always existed. Woman, 26. L. ptosis 7 years previously. Examination: Facial muscles flaccid. Muscles about mouth and eyes less exc. to elect. Ophthal. ext. Procopovici: Arch. f. Augenheilk., xxxiv., 1896, p. 34. Bilateral paralysis of VII. and VI. nerve. Muscles about angle of mouth retained and the orbicularis palp. Schmidt: Deutsche Zeitschr. f. Nervenheilk., x., 1897, p. 400. Paralysis bilateral of VII. and VI., also of XII. L. > R. Also absence of left pectoral major. The VII. nerve paralysis was complete.

UNCOMPLICATED CONGENITAL FACIAL PARALYSIS.

- Delprat: Weekb. v. het Nederlandsch. Tijdschr. vor. Geneek., November 29th, 1890, No. 22, p. 697. Cit. from Möbius. Not accepted by Möbius, but included by Kunn. Onset said to have been after an acute illness when 3 years old. Examined when 16. Right, weakness of muscles which elevate mouth, especially zygoid maj. Left, weakness of all muscles except corrugator supercillii, orbicularis palp. and the zygoid. Decreased elec. excitability. Left amblyopia.
- Henoch: Vorlesungen über Kinderheilk., 1897, ix. Auflage, p. 22. Boy, 10. Left VII. nerve paralysis, which had existed since birth; also paralysis of soft palate and deafness on left side. No condition in ear to account for deafness.
- Stephan: Révue de Med., 1888, July, p. 548. Abst. Bernhardt, Neurolog. Centralbl., ix., p. 423. Noticed soon

after birth. Examined when 32. Left VII. nerve paralyzed. Soft palate paralyzed. M. M. not excitable to elec. Taste normal. Deaf in left ear.

Schultze: *Neurolog. Centralbl.*, xi., 1892, No. 14, p. 425.

Noticed directly after birth. Examined when 4. Total left VII. nerve paralyzed. Pupils $L > R$. Slight nystagmus in lateral position. Elec. examination, strong current causes contraction in left orbicularis oris, and nothing else on left side. This muscle is not noted as acting normally. No other abnormality.

Bernhardt: *Neurolog. Centralbl.*, 1894, xiii., p. 2. No-

ticed 2 weeks after birth. Examined when 24. R. VII. nerve paralyzed (forehead, eye and upper lip). Muscles of under lip and chin retained. Only the active muscles respond to elec. from nerve. Slight nystagmus in lateral position. Taste normal.

Kunn: *Beiträge zur Augenheilk.*, Heft xix., 1895, p. 1.

Case doubtful. Noticed just after birth. Examined at 16. Abnormal development of right side of face. Eyes and forehead normal. Below, marked asymmetry of face. Masseters and bones less developed. Weakness but no actual paralysis of VII. and V. nerves. Normal electrical response. Taste normal.

Bernhardt: *Neurolog. Centralbl.*, xvi., 1897, p. 296. No-

ticed in first days of life. Examined when 7. Face, R. VII. nerve paralysis. Muscles of lower lid acted, and she could pucker her lips as in whistling. Elect. examination: Marked decreased excitability in orbic. oris to direct current. Is excitable from other side. Nothing from nerve. Taste could not be tested.

Procopovici: *Arch. f. Augenheilk.*, xxxiv., 1896, p. 34.

Case I. Since birth. Examined at 18. Bilateral paralysis of orbic. oculi and frontalis. No other abnormality.

Case II. Noticed soon after birth. Examined at 34. Bilateral weakness of VII. nerve, R. more than L. Eye muscles normal. Hearing normal.

DISCUSSION.

Dr. Wm. G. Spiller referred to the relationship between the condition described by Dr. Thomas and progressive muscular dystrophy of the Landouzy-Dejerine type. In the latter affection the muscles about the eyes and mouth are much involved, and the disease may exist for years with the atrophy limited to the muscles of the face. In the patients whom Dr. Spiller had seen, one might easily believe, at first sight, that the seventh nerves were affected. The existence of the disease in Dr. Thomas' cases, in two members of the same family, was suggestive of progressive muscular dystrophy, though the speaker did not intend to make a diagnosis different from that formed by Dr. Thomas.

Dr. B. Sachs said that while listening to Dr. Thomas' paper, it occurred to him that the cases reported were possibly instances of progressive muscular dystrophy of the Landouzy-Dejerine type. If the condition could be traced back to birth, however, that would militate against the latter diagnosis. In one of the photographs shown by Dr. Thomas it appeared that the eyelids could be very nearly closed, if not completely so, which would lead one to entertain the idea that the muscular element was very much more at fault than the neural. The speaker said he was inclined to believe that in these various forms of congenital defective development, the defect was not observed merely in the neural part of the motor tract, but that other parts of the body were also poorly developed. In one case which he had reported, a defect of the pectoralis major muscle was found in connection with a distinct defect of the scapula. It was probable, he thought, that these cases were not necessarily neural in origin; while some of them might be, others showed that the muscular or osseous system alone was involved.

Dr. Hugh T. Patrick inquired whether an electrical examination of the eighth nerve was made?

Dr. B. Onuf asked whether Dr. Thomas attributed any etiological importance to maternal impression in these cases.

Dr. Thomas, in closing, said he did not believe that maternal impression had anything to do with the disease in the cases he had reported. It might be of interest, however, to state that in one case in the literature where a child was born with a facial defect, the mother, during her pregnancy, had made frequent visits to a physician, who was treating her mother with electricity for a facial paralysis.

Dr. Thomas said that the question of a probable relationship between this condition and progressive muscular dystrophy had been brought up before, and while the anatomical

changes in the muscles in certain cases of congenital ptosis were quite similar to those found in this disease, this did not, by any means, demonstrate that the cause of the atrophy was the same. In his cases the defects of development were not confined to the muscles of the face; there was also a defect of the ear and a defect in the hearing, and on this account he regarded the symptoms as congenital. In reply to Dr. Patrick, the speaker said he did not make an electrical test of the eighth nerve. In conclusion he expressed the opinion that it would be unusual to see a case of muscular dystrophy of the facial type which dated from birth and persisted until the twentieth year, in which only the muscles of the face were affected.

190. LA MALADIE DU SOMMEIL ET SON BACILLE (The Sleeping Sickness and its Bacillus). Cagigal and Lepierre (*La Médecine Moderne*, 9, 1898, p. 60).

Cagigal and Lepierre, the authors, observed a negro, 16 years of age, a native of Angola, who suffered with the sleeping sickness for three years. He was in the hospital of Coimbre more than two months, during which time there was an elevation of temperature, with remissions, up to some weeks before death, but during this latter period the temperature was constantly subnormal. The only physical peculiarity further was constant passage of ammoniacal urine with an excess of phosphates and relative azoturia. The examination of the blood showed a constant presence of bacilli and spores. The cultures of blood from the arm and hand on serum, gélose, gelatine, bouillon and peptone resulted as follows: At moderate temperatures most of the media remained sterile, except those of serum and gelatine, which produced homogeneous colonies, the serum after three days, the gelatine only after four weeks. With the culture upon serum the same results were obtained as with the blood. There was some difficulty in development of the microbe upon the culture media except with serum, a point upon which the authors insist. The microbe observed in the blood and cultures is a straight bacillus, sometimes incurved, a little larger at the extremities than at the centre; it makes filaments, it is very little mobile, takes anilin stains well, does not color with Gram, and appears not to have any processes; free spores were observed both in the cultures and in the interior of the microbes. The general aspect of the preparations resembles those of the anthrax bacillus; the growth is best at a temperature of from 30 to 37 degrees C.; a moist heat of 75 degrees C. kills it in a minute; it is a true aerobic bacillus.

Postmortem—Some microbes were found in the intraperitoneal fluid. Inoculations of rabbits and guinea pigs produced certain effects not unlike those observed in the human subject—identical temperature-curves, sub-normal temperature preceding death, loss of weight, an appearance of depression, feebleness, especially of the hind quarters, ammoniacal urine, and death in from twenty-five to fifty days.

MITCHELL.