ON SO-CALLED FACIAL HEMIHYPERTROPHY.

BY H. MACKAY, M.D., DEVIZES.

THE extreme rarity of this condition is a sufficient reason for recording a fresh case, and for recapitulating what is known of it.

Described first by Beck in 1836 (1) it was nearly thirty years before a second case was reported by Friedreich (2). The Heidelberg professor described a third case observed by Heumann (3), and to these, Passauer in 1866 (4) added a fourth.

In 1869 Trelat and Monod (5) published a monograph on the condition of unilateral hypertrophy, partial or complete. The authors collected twelve cases of this condition, some of which, viz., those of Devouges (6), Ollier (7), and Monod (8), have been referred to since as instances of facial hemihypertrophy. I have not included them in the present series of cases, however, because the original observers state regarding each case that the facial asymmetry was insignificant, or that it eluded measurement.

The next contribution of importance was that of Fischer, on "Giant Growth," in 1879 (9). Fischer reported a fresh case of facial hemihypertrophy under his own care, and endeavoured to classify the various cases of hypertrophy that had already been reported.

He distinguished between "true" and "false" hypertrophy, limiting the former term to those instances in which all the tissues of a part were equally enlarged, and the part remained functionally useful, and assigning the latter to those in which the enlargement was due to, and was restricted to, over development of one tissue only.

The division was unsatisfactory and is now obsolete. It left unclassified the cases in which more than one tissue,

e.g., fat, sebaceous glands, and connective tissue have undergone hyperplasia, but in which all the tissues of the part, *e.g.*, the bones, have not participated.

While undoubtedly the conditions of lymphangioma and telangiectasis which Fischer appears to have had in view in framing his distinction, ought to be excluded from the list of hypertrophies, yet the latter term has, by the common consent of pathologists, been applied to those cases where hypertrophy or hyperplasia of one tissue, such as connective tissue or muscle, has taken place without exhibiting the intrinsic characters of tumour formation.¹

Passing over the contributions of Ziehl (10), Schieck (11), and Finlayson (12), each of whom described a fresh case, Lewin (13) in 1884 published an elaborate paper on what he considered to be the correlated conditions of facial hemiatrophy, facial and bodily hemihypertrophy, and cutaneous pigmentation. He gave a list of published cases in each class, including a case of facial hypertrophy of his own. He included in the latter condition, however, two cases published by Stilling (14), to which as cases of facial hemihypertrophy exception may reasonably be taken. Both were cases of swelling of the soft parts of the cheek, in one instance as large as a child's head, in the other as large as a man's fist, which appeared subsequently to injuries in this region. Both were accompanied by extreme dilatation of the blood-vessels of the part; in one case telangiectatic excrescences the size of raspberries being present, while in the other a semi-erectile condition existed, the swelling becoming plainly larger and more distended when the patient bent over.

Inasmuch as local ectases and vascular dilatations are of frequent occurrence in the records of undoubted cases of

¹ Whether osseous hypertrophy—hyperostosis, pure and simple—of a part such as the face or skull should be placed in a category apart from the condition of mixed overgrowth of bones and soft parts, which, in the majority of instances characterises facial hemihypertrophy, appears to be at present undecided.

Mr. Hutchinson has described (26) as "Unilateral Hypertrophy of the Skull" a case, to be referred to later, which would probably be classified during life as unilateral facial (and cranial) hypertrophy.

Brissaud also (62) has recently described as "hemicraniosis" two cases in which the condition is apparently identical with that described by Mr. Hutchinson. To these cases also reference will be made later.

facial hypertrophy, and it is difficult to decide from Stilling's account how much of the swelling in each case was due to angiomatous conditions and how much to hyperplasia of tissues other than vascular, I have not felt justified in altogether omitting these cases, but have placed them, as doubtful instances, in a division apart.

It is, however, to the Salpêtrière school that we owe the most important recent studies of the subject. Sabrazés and Cabannes (15) in 1898 described a fresh case with a bibliography. Their list of published observations, however, requires revision, as they have included as instances of facial hypertrophy, cases (*e.g.*, *Obs.* xiii. in their series) in which the face was not affected. These authors distinguish between the congenital and the acquired cases, and assign to each a different pathogeny. To this point reference will be made later.

Still later, the teachings of Brissaud on the influence of metamerism in nervous disease, and of Meige on chronic dystrophy, have drawn the attention of many of their pupils to the subject of hypertrophy of limbs, and other portions of the body. Unilateral hypertrophy of the face has also come within the purview of these studies.

The question whether facial hypertrophy, in any form, may be considered as a dystrophy allied in its nature to trophœdema or not is yet an open one. Meige himself (16) adopts towards the suggestion an attitude of caution.

As regards several instances of facial hemihypertrophy, however, *e.g.*, those of Beck, Berger, and Ziehl, in which the facial enlargement has concerned the soft parts only, and has exhibited the characters of a swelling, not pitting on pressure, colourless, and painless, it would appear to be difficult to separate the condition clinically from that described as "chronic trophœdema" of Meige. It is for this reason that I have included in the list of cases given below, the examples described by Rapin (17), Hertoghe (18), and Thomas (19).

English and American observations of the disease have been scanty.

Barwell (20) described a case in 1875, and again in 1881.

Heath (21) figured a case in "Injuries and Diseases of the Jaws." Sir Geo. Humphrey (22) described and figured a doubtful case, in which the gums were chiefly concerned, in 1886. In each of these cases it was the surgical rather than the neurological interest of the case that appealed to the observer.

Dr. Montgomery, of San Francisco (23), in describing a case, published a brief but excellent résumé of the subject in 1893.

Mr. Hutchinson and Dr. Alexis Thompson (26) have each reported cases of what the former describes as "Unilateral Hypertrophy of the Skull," and have admirably described, from skulls in their possession the osteo-pathic changes which occur. As regards the facial condition of these patients during life, the details are not sufficiently full to allow of their being tabulated with the other cases here presented. While it is in doubt, however, whether and to what extent the soft parts were involved, I think there need be no hesitation in including them as cases of facial (and cranial) hemihypertrophy.

Brissaud and Lereboullet (62), in the cases already alluded to above, describe a condition of hyperostosis of one half of the face and skull in the region of the trigeminal nerve, for which they propose the distinctive title of "hemio-craniosis."

In the first case epileptic attacks commenced at the age of 20 in a patient in whom, from the age of two months, there had been noticed asymmetry of the face, with bossing of the left frontal bone. Headache, but no vomiting, was present, and the fundus was normal.

In the second, the enlargement of the face and skull had also existed from childhood. The right half of the face was hypertrophied, and there was a well-marked painless boss in the right frontal region. Latterly, there ensued vomiting, headache, strabismus, double choked disc, exophthalmos, and ultimately complete blindness, leading up to exitus lethalis.

The autopsy showed the presence of numerous small angiolithic tumours on the inner surface of the dura on the

right side, and multiple exostoses, the size of nuts, on the inner aspect of the cranium on the same side, which corresponded to correlated depressions in the cerebral substance.

The authors claim for these cases a certain relationship to facial hemiatrophy, inasmuch as in each condition there is present a unilateral trophic affection in the region of the trigeminal.

The condition thus depicted appears to be identical with that described by Hutchinson and Alexis Thompson as unilateral hypertrophy of the skull, and by other observers as facial hemihypertrophy, but, in deference to the distinctive nomenclature adopted for it by so distinguished an authority as Brissaud, I have excluded these observations from the subjoined list of cases of facial hypertrophy.

In the cases here given, the original records have been consulted in all instances, with the exception of Cases 1, 14, and 22.

In addition to the observations of Ollier, and Monod, already referred to, I have omitted several others included by former writers, on account of the poverty of detail in the available records. Such are those of Steffen (27), Zhukovsky (28), and Stembo (29), quoted by Messrs. Sabrazès and Cabannes, and that of Logan given in the *Virchow-Hirsch Jahresbericht* for 1868.

Of the associated, but commoner, condition of unilateral hypertrophy of the body, the literature is too voluminous to attempt to refer to it here.

Case 1.—BECK. *Medicinische Annalen von Puchelt, Chelius und Naegele*, 1836, Bd. II., No. 1, p. 89 (quoted by Ziehl—*Virchow's Archives*, Bd. xci.).

A girl of 3 years presented a hypertrophic fulness of the right cheek, which had been present since birth, and increased with the growth of the child. The soft parts alone were affected.

Case 2.—FRIEDREICH, 1863 (quoted by Ziehl), *Virchow's Archiv.*, Bd. xxviii., p. 474.

A girl of 16 showed soon after birth enlargement of the right side of the face, chiefly marked in the infraorbital portion. The forehead and eyes were symmetrical. The external ear was

enlarged in all directions. The hypertrophied side showed circumscribed patches of pigmentation, and an enormous development of abnormal hairs existed on the ear, cheek, temple, and along the jaws. The hairy scalp and the hands showed no abnormal development in the hairs. The irides showed similar coloration. An exaggerated sebaceous secretion covered the neighbourhood of the ear and mastoid with a greasy layer.

On the right side the alveolar processes of the upper and lower jaw were double the thickness of the opposite side; the teeth also were larger, longer, and thicker than those on the left. Tooth-ache was frequent.

The right tonsil and right half of the tongue were much enlarged, and the saliva was more abundant on this side. On the hypertrophied half of the tongue sensibility to taste and touch were less developed. The right cheek was of a deeper colour than the left, and experienced a subjective sensation of warmth. The child suffered from otorrhœa on the right side.

An autopsy was obtained, but nothing throwing any light on the condition was found. The nerve trunks at the base of the brain were unaltered (macroscopically). The petrous portion on the right side was hypertrophied, and showed exostoses projecting from its surface which had indented and compressed the opposing surface of the brain.

Friedreich also communicated the following case:—

Case 3.—HEUMANN (*loc. cit.*).

A girl, five years old, had shown since birth enlargement of the left side of the face, which involved the cheek, the two jaws, the teeth and the tongue. The skin of the ear was thickened, and the hairs more developed. There had been a fœtid otorrhœa from the left ear since birth, but the hearing was not impaired. There was a constant flow of saliva from the left side of the mouth.

The child died from meningitis following caries of the petrous bone. No autopsy.

Case 4.—PASSAUER, 1866, *Virch. Archiv.*, xxxvii., p. 410.

A boy of 11 showed congenital hyperplasia of the left side of the face. During pregnancy the mother was crushed by a wagon against a wall, and suffered severe abdominal pain for some time.

The enlargement involved the cheek, upper and lower lips, and the chin, and was limited by the median line of the face. The

alveolar borders of the upper and lower jaws were much enlarged on the left side, as also was the left half of the tongue and palate, and the teeth. On the hypertrophied portion of the face there was no play of expression.

Case 5.—BERGER. "Zur Aetiologie und Pathologie der sogenannten Muskelhypertrophie." *Deutsche Archiv. für Klin. Medicin*, 1872, p. 393.

In a woman of 58, Berger noticed hypertrophy of the face supervene in the course of severe and obstinate trigeminal neuralgia. The hypertrophy concerned the soft parts only, and especially the subcutaneous connective tissue.

Case 6.—FISCHER. "Der Riesenwuchs." *Deutsche Zeitschrift für Chirurgie*, Bd. xii., 1879.

In a boy of 7 months the left side of the face had been hypertrophied since birth. The enlargement and thickening affected all the tissues. The skin showed normal structure, and the bones were smooth. Of the latter those chiefly affected were the upper jaw and the frontal bone. The circumference of the head in the frontal plane was 56 cm., of which the left half measured 31, and the right 25 cm. The tongue was normal, and the hairs of the scalp, eyebrows, and eyelashes were equally developed on both sides. There was no difference in the eyes, or in the lachrymal secretion of the two sides. No discolouration was present. Motility, sensibility and temperature, were normal.

Case 7.—BARWELL. *Transactions of Clinical Society*, vol. xxxii., 1881.

A boy of 13 showed enormous hypertrophy of the right side of the face. It affected the head and face. (It appears to be chiefly infraorbital in the illustration. H.M.)

The texture was soft and yielding.

Near the ear was a patch of dark horny ichthyosis (true hypertrophy of the cuticle), which was increasing in size.

The upper and lower maxillæ were massive and thick, and the teeth were hypertrophied also. The tongue was enlarged on the affected side.

Mr. Barwell suggests a perverted action of the sympathetic as a probable cause. The common carotid was tied, and the patient died from secondary hæmorrhage.

As an appendix to the above may be quoted a previous observation of Mr. Barwell's recorded in the *Medical Times and*

Gazette, vol. i., 1875, which, though not unilateral, possesses considerable interest. The patient was a lad of 19, who showed hypertrophy of both sides of the face. It affected the soft parts only, the bones being of natural size and shape. It was stated to have appeared in a single night.

Mr. Barwell tied both facial arteries, but without much improvement.

Case 8.—ZIEHL. Virchow Archiv., Bd. xci., s. 92, 1883.

A boy of 4 years presented hypertrophy of the right side of the face which had existed since birth. Four or five weeks before her confinement the mother had been stung by nettles on the right side of the face. At five weeks old the child was convulsed, due to hydrocephalus. Subsequently fits were frequent. The enlargement affected the lower half of the right side of the face. The brow, skull, eyebrows, eyelids, and lashes were unaffected, but the hair on the right side was stiffer and coarser than on the left. There was convergent squint of the right eye with proptthalmos. The right iris was larger than the left, and the right ear longer and broader. Brown pigment spots were present at many places in the face. The alveolar processes of the upper and lower jaw were enlarged, but the teeth were equal on both sides. The right half of the tongue and right tonsil were much enlarged, and there was increased salivary flow from the right side of the mouth.

Motility and sensibility normal. The right hand was larger than the left.

Case 9.—SCHIECK. "Ein Fall von einseitiger Gesichtshypertrophie," Berlin Klin. Wochenschrift, 1883, p. 200.

The patient was a girl 9 years old, born of a family with no neuropathic tendency. Nothing abnormal was noticed at birth.

At the end of the second year there appeared an increasing swelling of the left cheek and upper lip. At the same time further development of teeth in the upper jaw of the same side ceased. The child was obtuse mentally. At 9 years the overgrowth affected the soft parts of the left cheek and upper lip, and to a less degree the left lower lid. The left nostril was thicker than the right. The lower part of the face and the philtrum of the upper lip were pushed over to the right side.

The left cheek was redder, and the growth of hair distinctly stronger. Sweat and sebaceous secretion not increased. Of the

bones the zygoma, the body of the left maxilla, and particularly the left upper alveolar process were more strongly developed than the right. The left upper jaw contained only the two incisors, canine and first premolar. The lower jaw presented nothing abnormal.

The left half of tongue and left tonsil not enlarged. Left ear and brow normal. Growth of hair on both sides of skull equal, as also pupils. Motility and sensibility normal.

Case 10.—EVE. Transactions Pathological Society, vol. xxxiv., 1883.

A woman of 34 had congenital hypertrophy of the foot and leg on the left side, which had rapidly increased.

The left half of the head and face were also larger, as was the left half of the tongue.

The autopsy showed symmetrical enlargement of the left side of the brain, with several osseous growths, the size of peas, in the dura. A fibrous tumour, the size of a filbert, grew from the outer side of the left cornea, and an epulis, the size of a pigeon's egg, from the left upper jaw. The left patella showed hyperostosis.

Case 11.—FINLAYSON. "Case of a child affected with congenital unilateral hypertrophy. — Glasgow Medical Journal, November, 1884.

A female child of 18 months had suffered from repeated attacks of otorrhœa in the right ear. From birth the right side had been hypertrophied throughout. The facial enlargement affected the bones as well as the soft parts. The parietal protuberance was excessively developed, as also were the upper and lower jaws. The tongue on right side and right tonsil were larger. The palate and uvula were normal. The temperature on the right side of the face was 3° to 4° higher than on the left. Teething was precocious on the right side.

On the left side the index, middle, and ring fingers were hypertrophied. Most excessive, however, in size was the middle finger of the right hand.

Under the excitement of crying, patches of colour, red, purple, or blue, come out over the skin in the vicinity of the navel, on the inner and posterior aspects of the hypertrophied right leg, and on the back. The patches suggest cutaneous nævi. The discolouration fades as the child becomes quiet.

Case 12.—LEWIN. "Studien über die bei halbseitigen Atrophien und Hypertrophien, namentlich der Gesichts vorkommenden Erscheinungen, &c."—*Charité Annalen*, Bd. ix. s 619, 1884.

A man of 26 showed crossed hypertrophy, affecting, on the left side, the face, neck, and arm, and on the right side the leg, which was longer and thicker than the left.

In the face the hypertrophy concerns the soft parts, and to a less extent the bones, viz., the frontal, zygoma, and upper and lower jaws. The hypertrophy is limited, vertically by the middle line, and otherwise by a line from the middle of the brow to the upper insertion of the auricle.

The skin over this area is hyperæmic, of a bright red colour, and hyperidrotic. The conjunctival and pharyngeal mucous membrane are also hyperæmic, and so is the left half of the hard and soft palate. Tongue and teeth normal. Hyperæmic patches are present on the chest, and flecks of vitiligo are present on the left half of the abdomen, reaching to the middle line, but not passing beyond it. Nævi melanoides are present on the left side of the neck. The sensibility to temperature is diminished on the right side, and general sensibility on the right side of the tongue only.

Left-sided toothache had been frequent since childhood.

The temperature was lower by $\frac{6}{10}^{\circ}$ C. in the left axilla, though the patient had often a subjective sense of warmth on this side.

Case 13.—HUMPHREY. "Annals of Surgery," vol. iii., 1886.

A boy of 12 suffered from unilateral hypertrophy of the gums of the lower jaw on the left side, which were thicker than those on the right.

There was no discoverable difference between the bones of the two sides of the head and face, but on the left side the tonsil and soft palate were larger than on the right. The upper and lower lip on the left side, protruded and everted, were thick and coarse. The left eyelid was larger and the eyelashes longer than the right, and the hairs of the left eyebrow were coarser and more numerous. The pinna of the left ear was larger and thicker than the right. The hair of the scalp came down lower in front of the ear, and the skin of the face was fuller and more covered with hair. The papillæ on the left side of the tongue were also larger.

The second digit of the left foot and the right thumb were abnormal, ending in soft bulbous extremities.

Case 14.—MASMÉJEAN. "Thèse de Montpellier," 1887-8 (quoted by Sabrazés and Cabannes).

An infant of 13 months had hypertrophy of the right side of the face, and of the limbs on the right side. The hypertrophy did not affect the length of the limbs, but only their thickness. The augmentation appeared to be chiefly due to an increase of fatty tissue.

Case 15.—KIWULL. "Ein Beitrag für Casinstik der halbseitigen Gesichts-hypertrophie." *Fortschritte der Medicin*, 1890, Bd. viii.

A girl of 18 showed right-sided hypertrophy of the face, which was congenital, and progressive. It affected both the bones and the soft parts.

There was bossing of the right frontal bone and prominence of the supraorbital ridge. The upper and lower jaws were enlarged and the surface of the latter was irregular and rough. The right alveolar processes were twice as large as the left. The teeth were widely spaced apart, the lips greatly thickened, and the ear enlarged. A zone of hyperæmia extended obliquely to the right from the middle of the nose for a distance of 2 cm. The right half of the tongue was three times its normal size, and the right tonsil also enlarged. From the right angle of the mouth ran a constant flow of saliva. Taste was blunted on the right side of the tongue, but sensibility of the face was unaltered. Portions of the skin and lip were removed for cosmetic reasons.

On examination, the various layers of the epidermis and cutis were of normal thickness. The sebaceous glands were hypertrophied, and the hair follicles normal. The fatty and connective tissues were increased in bulk, the former beneath the skin, and the latter beneath the mucosa. This latter, made up of fine fibres with few cells, was traversed by dilated veins and arteries. The muscle fibres took but slight part in the overgrowth.

Case 16.—TILANUS. *Münchener Medicinische Wochenschrift*, No. 4, 1893.

A girl of 10 had hypertrophy of the right side of the body, including the face. It was not noticed at birth, nor till the child was 3 years old. It had increased considerably of late.

The facial enlargement concerned the cheek, which was thicker and wider, and the right half of the tongue. The cranial circumference was also increased on the right side. The right shoulder, arm, half of trunk, buttock, leg and foot, all showed overgrowth. The musculature on this side was more powerful.

Electrical reactions and reflexes normal. There was a reticulated marbling of the skin of the trunk and limbs. Under the influence of cold the colour of the marbling became darker, and even blue.

The author thinks it possible that a central cause may be at the root of the overgrowth, and refers to the case of Demme.

Demme's case (*Wiener Medicinische Wochenschrift*, No. 2, Jahrgang xiv., 1891) was one of congenital unilateral hypertrophy in a baby girl who lived only a few days. The hypertrophy affected the whole of the left side of the body. On autopsy the cross sections of the cerebral hemisphere on the left side were larger than those on the right. The muscle fibres on the left side were hypertrophied on cross section, and the nuclei of the sarcolemma increased in number, and arranged in rows.

Case 17.—MONTGOMERY. Medical News, vol. ii., 1893, p. 61.

The patient was a man of 31, of German extraction. At 2 years he had brain fever. At 8 or 9 an alveolar abscess formed on the left lower jaw and caused much pain. No neuralgia. No abnormality of dentition.

A photograph taken at 5 shows the face to be perfectly symmetrical.

At 10 the gums were noticed to be enlarged, and at 12 the deformity was noticeable on the left side of the face.

At 31 the deformity occupied an irregular quadrilateral enclosed internally, by the mesial line from the anterior extremity of the sagittal suture, down to the mental symphysis, below by the border of the lower jaw and on to the tip of the mastoid, and posteriorly from the mastoid to the sagittal suture. The hypertrophy is most marked in the supraorbital, molar, and mental regions, where the tissues are raised up into tumefied masses consisting of soft parts and bone. The frontal bone shares in the hypertrophy, a mass of bone running upwards and backwards over this region from the outer extremity of the supra-orbital ridge.

The scalp and hair are normal. The upper half of the left ear is hypertrophied. The discs are excavated, and not engorged. The left lower jaw is enlarged and roughened. No enlargement of teeth, tongue, or tonsil. No sensory abnormality, general or special. The deformity is progressive.

Portions of skin removed for æsthetic purposes showed, on section, increase in the size of the hairs and sebaceous glands. Also plugging of the sebaceous glands with sebum, and degenerated cells. There was no increase in connective tissue, or fat.

Case 18.—DANA. *Journal of Nervous and Mental Disease*, November, 1893, page 234.

The patient was a professional giant of 19. Height 7 ft. 4 in., weight 325 lbs.

Acromegaly was present, the hands and feet being enormous. He "wore a shoe just a foot and a half long." The enlargement was symmetrical in both upper and lower extremities. The left half of the face was hypertrophied, the enlargement having begun at puberty, and being progressive. It corresponded closely with the distribution of the left trigeminus, and involved the upper and lower jaw, the alveolar processes being enormously enlarged, as also the palatal arch. The teeth were normal, and so were the orbits, but the left frontal bone showed great bulging.

No pigmentation. Muscular system moderately developed, but the patient was weak. Vision good. No contraction of fields. Pupils normal. Intelligence good. Appetite prodigious.

Case 19.—HEATH. "Injuries and Diseases of Jaws," 1894, page 136.

A boy of 12 showed enlargement of the face, which was noticed at three months old. It affected the bones and soft parts on the left side.

The superior maxilla on this side retained in its alveoli the temporary incisors and canines, but these had been replaced by permanent teeth on the sound side.

Mr. Heath removed the superior maxilla, but the patient's condition, two years later, was as unsightly as before.

Case 20.—ARNHEIM. "Hypertrophie congénitale de la moitié droite de la face et du bras correspondant."—*Semaine Médicale*, 1895 and 1897.

A girl of 4 months exhibited congenital hypertrophy of the right side of the face and the corresponding arm. The hyperplasia of the face formed a prominent mass, which raised the left lip.

The right half of the tongue was enormously hypertrophied, and the right ear was larger by 1 cm. than the left. The right arm also longer than the left.

Dyspnoea was present, of cardiac origin.

At 2 years the child died of broncho-pneumonia. The autopsy showed that the hypertrophy extended to the internal organs on the right side of the chest. The right auricle and ventricle were larger than the left, and the right bronchi more spacious.

Case 21.—SABRAZÈS and CABANNES. “Hemi-hypertrophie faciale.”—*Nouvelle Iconographie de la Salpêtrière*, tome xi., 1898.

A boy of 3 years presented congenital hypertrophy of the right side of the face.

At the fourth month of pregnancy the mother suffered a fall on the abdomen, and at the seventh an emotional shock. At four months the child had a purulent otorrhœa (right), which lasted two months, and left impaired hearing. Dentition was precocious on this (right) side. The original disproportion has been maintained since birth.

It concerns the bones and soft parts, and the frontal boss is enlarged. The cranium is asymmetrical, showing increased development and fulness on the right side. Venous dilatations are present on the mucous surfaces of the left cheek.

In the median line in the occipital region there is present in the scalp a pigmented streak, café-au-lait colour, extending from the nape of the neck to four fingers' breadth below the summit of the vertex; over this streak the hair exhibits a negroid curliness.

Electrical examination shows on the left a normal reaction, and on the right a diminution of excitability, owing to the increased resistance of the tissues.

Sensibility to pain is diminished slightly on the affected cheek. On the forehead it is the same as on the opposite side. The gustatory sense is normal. The pharyngeal and palpebral reflexes are normal.

The right cheek always redder than the left, becomes scarlet when the patient approaches the fire. In winter it is more sensitive to cold, and suffers from cracks.

The right hand is slightly larger than the left.

Case 22.—HERMANIDES. “Een geval van hemi-hypertrophia congenita.”—*Psychiatr. en neurol., Bladen*, 1899. (Abstract in *Mendel's Centralblatt*, 1900, p. 912.)

In a child of 13 months all parts of the left side of the body were larger than the right. The palpebral opening, eyeball, and pupil, appeared to be somewhat larger on the left side. As a rule the left ear was redder than the right. A striking difference obtained between the two halves of the tongue, but the small right half showed fibrillary twitchings, and the mucous membrane was thrown into small folds. Trophic signs were absent, apart from the increased reddening of the left ear. The teeth and hair on both sides were equally developed. Sensibility was equal on both sides, and no difference could be detected in the sensibility.

It appeared, at first, doubtful whether in this case the condition was one of left hypertrophy or of right hemiatrophy, but the author is satisfied that the former was the real condition.

Case 23.—HUTCHINSON. Polyclinic, July, 1901.

The patient was a middle-aged woman with unilateral hypertrophy of the face and skull. The soft parts did not appear excessively increased, but there was enlargement of most of the bones in the right half of the face. The frontal boss on the right side was specially prominent, and the right eye bulged forward, probably on account of a growth behind it.

There is no statement as to whether the condition was congenital or not.

Elsewhere the patient showed multiple cutaneous and subcutaneous fibromata scattered over the chest, and also osseous enlargement of the left side of the sternum, involving possibly some of the ribs.

Case 24.—RAPIN. "Sur une forme d'hypertrophie des membres (Dystrophie conjonctive myélopathique)." Nouvelle Iconographie de la Salpêtrière, No. 6, 1901.

The case was a congenital one, in a woman of 29. The hypertrophy concerned the right side of the face and the two upper limbs. In the face it affected the cheek and ear on the right side. The maxilla showed no thickening. The teeth were bad. There was no nævus or vascular taches.

The two arms were equally enlarged. The hypertrophy commenced about the middle of the limbs, but was most marked in the hands. The skin was white, soft to the touch and normal. There were no abnormal vessels on the surface. Under the influence of cold it became quickly cyanotic and stiff.

Palpation gave the impression of moderately firm fatty tissue. It did not pit on pressure. Cutaneous sensibility was normal.

Case 25.—HERTOGHE. "Contribution a l'étude de trophodémie chronique." Ibid., page 497, 1901.

A girl of 21 shows congenital hypertrophy of the right cheek. The tongue is not increased in size, but the papillæ on the affected side are hypertrophied. The teeth are more strongly developed on the affected side. On this side the wisdom tooth is through and well established, but not on the healthy side. The palate on the right side is larger, and its rugosities are more marked. The hairs on this side are hypertrophied.

The bones of the face are not affected, but the middle turbinate is hypertrophied as compared with the sound side.

A sister of the patient shows infantilism at 15 years, and has the lower limbs "en colonnes," the legs being as thick as the thighs. The mother suffered from lipoma of the arm.

Case 26.—THOMAS. "Deux cas d'hémihypertrophie congénitale du corps." *Ibid.*, page 508, 1901.

A female child of four months has congenital hypertrophy of the face and one half of the body. The hypertrophy affects the right cheek, which is plumper and thicker than the left. The skin also appears thicker. The palpebral aperture is wider on the right, but there is no anomaly in the eyes, ears, nose, or mouth. The right half of the tongue is much larger and thicker than the left. Palate and soft palate normal.

The bodily hypertrophy affects the trunk and limbs of the left side. The bones are enlarged as well as the soft parts, and the muscles also participate. The skin is thicker on the affected side.

The hypertrophy is more pronounced in the thigh than in the leg and foot. Coloration, temperature, and sensibility normal.

Two years later the child was examined again. The hypertrophy was still present, but was much less apparent. Teething was delayed, the child at this date having only six.

Case 27.—THOMPSON. *Transactions Med. Chirurg. Soc. of Edinburgh*, vol. xxi., page 204, 1901-02.

In a young woman of 27 enlargement of the left cheek, with hyperostosis of the left upper jaw and malar bone had existed, probably since birth.

The hyperostosis affected the entire upper jaw, except its alveolar margin, and the downward projection of the alveolar margin caused obliquity of the oral fissure. The nose deviated to the opposite side owing to overgrowth of the nasal process.

Case 28.—FOWLER. *Ibid.*, 1901-2.

A girl of 3 showed hemihypertrophy of the right side, affecting the face, arm, thorax, and leg. The distal parts were more hypertrophied than the proximal.

The temperature was higher on the affected side, and the eruption of teeth was earlier. Numerous capillary nævi were scattered over the body.

Case 29.—MACKAY. Unpublished.

A girl of 10 suffers from acquired hypertrophy of the left side of face and skull. The child was normal at birth, and a portrait taken at 5 shows no asymmetry of the face. Measles occurred at 8, followed by earache and otorrhœa, and the latter complaints have been frequent since. There has been no general headache, and no trigeminal neuralgia. Memory and intelligence are good. Bodily symmetry is elsewhere perfect, except that the left hand is somewhat larger than the right. The deformity was noticed first at 6 years of age as a bony enlargement on the head near the vertex. Then appeared a similar swelling in the occipital region. Then came a fulness in the forehead, and, lastly, the cheek began to enlarge.

FIG. 4.

Cast of upper dental arch, showing hypertrophy of left alveolar border and teeth. The hypertrophy is sharply defined by the mesial line.

The child is of average build and well nourished. Height 4 ft. 5 in., weight 68 lbs. The bony skeleton elsewhere than in the affected parts is normal. There is no sign of rickets or of congenital syphilis. The teeth are mostly carious. The facial enlargement is limited strictly by the middle line. The left cheek is fuller and thicker, also the left upper and lower lips. Left nasal bone is thickened. Soft parts of nose on left side hypertrophied, and also the left half of the chin.

The bony overgrowth extends to all the left facial bones, and

FIG. 1.

Hemihypertrophy of face and cranium.

FIG. 2.

Left side view, showing frontal and parietal bossing.

FIG. 3.

Normal side.

to most of the cranial, *e.g.*, the frontal, parietal, sphenoidal (orbital plate, probably), temporal and occipital.

There is marked bossing of the frontal, a smooth prominence of bone commencing at the inner end of the supraorbital ridge, and passing upwards over the middle of the bone to the hairy scalp, where it ends.

Another bony ridge commences to the left of the sagittal suture near the vertex, and runs downwards and forwards over the left parietal bone.

A third mass in the form of a rounded boss occupies the superior angle of the occipital bone posterior to the fontanelle. This boss is mesial in position. In the parotid region the enlargement of the malar and zygoma give rise to a fulness suggesting mumps. The superior maxilla is greatly enlarged, and also its alveolar border. The molars are longer and thicker on this side. Behind the ear the mastoid and the adjoining portion of the temporal bone are enlarged.

Exostoses project from the tip of the mastoid, and from the posterior free border of the superior maxilla.

The posterior part of the skull on the left side is also enlarged, and the hair covering this portion is coarser than on the right. Tongue, palate and tonsils are normal. In the left eye the sclerotic in the lower and outer quadrant of the ball is bulged forward, probably owing to some pressure from behind. The rim of the orbit is thickened. Owing to the pushing forward of the outer part of the ball an internal squint, not congenital, has been developed.

Ophthalmoscopic (left eye).—The media are clear, but there is oedematous swelling of the disc, with haziness of the margin, everywhere except in the lower nasal quadrant. The retinal vessels are obscured by the swelling. On the temporal side the retina is swollen and hazy.

Vision R. $\frac{6}{6}$, J.1 at 22cm.; no H.M.

L. $\frac{6}{36}$, J.20; not improved by glasses.

The F. of vision, normal in the right eye, is contracted in the left to the nasal side.

Hearing on the right side is normal, but on the left the watch-tick is heard on contact only. There is an old perforation of the drum.

The special senses are otherwise normal.

Ordinary sensibility to touch, pain, and temperature is unaffected.

Motility and reflexes are normal.

There are no vaso-motor signs, increased sweating, dilated vessels, flushing, or hypersensitiveness to cold. No salivation. There is, however, hypertrichosis of the left cheek in the neighborhood of the ear.

Electrical reactions normal.

Stilling's cases are briefly as follows: ("Untersuchungen über die spinal Irritation." Leipzig, 1840, S. 337).

(1) A man of 48 had been thrown when 23 years of age from his horse, injuring his face. Since then the cheek (right) had been larger than the other. Subsequently the same cheek was gored by the horn of an ox. At the time of examination the right cheek was the size of a small child's head. It was soft and smooth and not painful on palpation. At several places small telangiectases the size of raspberries were present. The skin was anæsthetic over an area which was sharply defined. Movement was also lost on the affected side. The tongue and oral mucous membrane were normal. So were the special senses. No pulsation, but the arteries collectively on the affected side, and apparently the carotid, too, were markedly enlarged.

(2) A woman of 38, when nine years old, was struck by a stone on the right cheek, and lost much blood from the mouth. A swelling gradually developed which resisted all treatment. It was the size of a man's fist and limited itself exactly to the right cheek. The skin felt soft, and had the same healthy red colour as the other side. The swollen mucous membrane protruded on the right angle of the mouth showing several enlarged dark coloured blood vessels. Much saliva escaped, which Stilling considered to be serous exudation from the enlarged blood vessels in the mucous membrane. The oral mucous membrane covering the cheek was of a dark blue-red colour, tumid and swollen, with numerous distended blood vessels.

Sensation, motility and reflexes normal. The facial bones were not affected.

When the patient held herself bent over, the swelling became larger and more distended.

The temperature was the same on both sides, and no pulsation was present.

DIGEST OF CLINICAL FEATURES.

Females are affected oftener than males, in the proportion of 17 to 12. The right side is hypertrophied oftener

than the left, in the proportion of 15 to 12. This accords with St. Hilaire's (63) opinion regarding unilateral hypertrophy of the body. There is no evidence of hereditary transmission. "Maternal impressions" figure on three occasions.

In twenty-three cases the condition was congenital, or was probably so. In six there is good evidence that it was post-natal, or "acquired." The latter form has appeared at ages varying from 6 to 55 years.

As regards personal antecedents, in four cases (Nos. 2, 3, 21, 29), there has been otorrhœa on the same side as the hypertrophy. In the former three, congenital cases, the otorrhœa could not well have preceded the overgrowth. In the latter case it did. In four other cases there has been present some peripheral irritation of the trigeminal nerve, from frequent toothache (Nos. 2, 12), or from obstinate neuralgia (No. 5), or alveolar abscess (No. 17). In the two former cases the temporal relation of the pain to the hypertrophy is not stated. In the two latter the pain preceded the overgrowth.

In the majority of cases, 19 out of 29, the hypertrophy has been limited to the face, or the face and head. In the remaining ten other parts of the body have been affected also. Of the nineteen former cases, in three (Nos. 1, 5, 25) the soft parts of the cheek were alone affected, in sixteen the facial bones shared in the hypertrophy. In eleven of these the cranial bones were also involved. In three additional cases (Nos. 20, 24, 26) the soft parts of the cheek were hypertrophied, but in conjunction with segmental overgrowth of some other part of the body.

In three cases (Dana's case, and the skulls belonging to Mr. Hutchinson and Dr. Thompson),¹ and also in the two cases of hemicraniosis recorded by Brissaud and Lereboullet, it is stated that the parts affected corresponded exactly to the distribution area of the fifth nerve. As a matter of fact, however, this correspondence with the trigeminal area, either in whole, or in part, appears in every recorded case of

¹ *Vide infra.*

facial hypertrophy. In one case only have the limits of the trigeminal been transgressed, viz., in case 29, where an occipital boss was present.

This relation of hypertrophy to peripheral nerve area is scarcely to be wondered at when we recall that, with the exception of the parotid region, supplied by a branch of the great auricular, the trigeminal supplies practically the anterior two thirds of the cephalon. In the light of Brissaud's (46) observations on the influence of metamerism in nervous disease it is interesting to note that according to Gegenbauer (40), the trigeminal though a single peripheral nerve represents the complete sensory fibres appertaining to the first complete cranial metamere.

As regards the bones, those most frequently hypertrophied were the maxillæ, one or both being enlarged, either in part, or wholly, in 17 cases out of 29. When, as happened in three cases, one maxilla only was affected, it was the upper one that was selected.

In connection with this preference for the maxillary bones it is noteworthy that, as Professor Dixon has pointed out,¹ a similar selection takes place in acromegaly. Professor Dixon calls attention to the fact that the pituitary body is developed partly from the neural tube, and partly from the buccal cavity, and that when this body is diseased, as in acromegaly, the first bones to be enlarged or affected are those developed as membranous bones in connection with the primitive mouth, or stomodæum, *i.e.*, portions of the superior and inferior maxillæ.

Of the cranial bones the one most frequently affected was the frontal, which displayed bossing in six cases. In two more the parietal also showed enlargement, and in one (No. 29) the occipital showed a mesial boss. In three cases there was uniform unilateral enlargement of the cranial bones, without bossing. Thus in 11 cases out of 29 the osseous hypertrophy was hemicephalic rather than hemifacial.

Coming now to the cases in which the trunk and

¹British Medical Association Meeting at Oxford, 1904. Discussion on Professor Cunningham's paper. Anatomical section, July 27.

appendages shared in the overgrowth, in five of the ten observations the hypertrophy was unilateral, affecting the limbs or the limbs and trunk on the same side as the face (Nos. 11, 14, 16, 22, 28). In one (No. 20) the upper limb only, on the same side, was affected.

In two the distribution was crossed, affecting either the face and arms on one side, and the leg on the other (No. 12), or the face on one side and half the body on the other (No. 26). In one (No. 24) the lower segments of the two upper extremities shared in the hyperplasia, along with half the face, and in one (No. 18) the facial hemihypertrophy was associated with acromegaly and giantism.

In addition to the cases already mentioned where the distribution of the hypertrophy in the appendages was segmental—*i.e.*, was limited by a boundary line, running at right angles to the long axis of the limb—the same feature was present in Case 26, where the thigh showed more marked enlargement than the leg and foot of the same side, and in Cases 21 and 29 where one hand, on the same side as the face, was enlarged in each case (cheiromegaly). A segmental distribution of the hypertrophy of the appendages was thus present in seven cases out of twenty-nine.

Unilateral enlargement of the tongue occurred in eleven cases. In six the corresponding tonsil was enlarged, and in five the teeth shared in the hypertrophy.

Cutaneous pigmentations, nævi or marbling of the skin, were present in seven cases. In one (No. 7) there was a patch of dark horny ichthyosis near the ear, and in one (No. 12) spots of vitiligo were scattered over the abdomen, on the same side as the hypertrophy, their distribution being bounded by the middle line.

Vaso-motor, or sympathetic, disturbances, such as hyperidrosis, excess of saliva, cutaneous hyperæmias, localised elevations of temperature, and excess of sebaceous secretion, are noted in seven cases (Nos. 2, 8, 9, 11, 12, 15 and 21).

Coarseness, or abnormal growth of hair, a trophic, probably, rather than a vaso-motor, symptom is reported in six cases (Nos. 2, 3, 8, 9, 13, 29).

As regards nerve function, motility and general sensi-

bility are usually stated to be normal, but the latter was diminished in two cases (Nos. 12, 21). Subjective sensations of warmth, or a special sensitiveness to cold, with a cyanotic change of colour, under its influence is reported in five cases (Nos. 2, 12, 21, 16, 24).

Dissociation of sensation does not appear to have been looked for, unfortunately, in the majority of the recorded cases. In Lewin's case (No. 12) it was present.

The special senses were but seldom affected.

In two taste was blunted, and in one (No. 29) sight and hearing were damaged on the affected side. The defect of sight in this case was associated with neuro-retinitis, apparently due to pressure by osteophytic growth on the back of the ball, and the deafness was connected with chronic otorrhœa.

Prophthalmos was present in three cases (Nos. 8, 23, 29). The reflexes and electrical reactions were generally normal, and intelligence was usually unaffected.

In five cases the hypertrophy was progressive to a greater degree than the bodily growth (Nos. 1, 15, 17, 18, 29). In one (No. 26) it was retrogressive. This case, however, was an instance of the so-called chronic trophœdema.

Although, following the current nomenclature, the cases described here are distinguished as "congenital" and "acquired," it may be questioned whether the distinction is pathologically a valid one. That the condition is in the biological sense always acquired is evident by the fact that its hereditary transmission is unknown.

Even on clinical grounds it appears to be impossible—apart from the mere date of commencement—to separate between the two classes. Sabrazés and Cabannes are of opinion that a distinguishing feature may perhaps be found in the date of the dental evolution, which they consider to be delayed in the "acquired," and precocious in the "congenital, cases." But in the cases of Heath and Thomas (Nos. 19 and 26), which were "congenital," the dental evolution was retarded. Again, the unilateral enlargement of the tongue, which these authors consider to be usually absent in "acquired" cases, was present in that of Tilanus (No. 16), which was "acquired."

It does not appear possible, therefore, either on clinical or biological grounds, to maintain the distinction, and it would be more logical to consider the morbid conditions in both classes to be the outcome of some disease process common to them both, the date of commencement having been prenatal in one class and postnatal in the other.

PATHOLOGY.

Post-mortem observations of the condition may be said to be non-existent. In no case have the nerve centres been examined by modern methods.

Portions of soft tissues removed during life have shown as histological conditions, overgrowth of sebaceous and sudoriparous glands, and of adipose and connective tissues, with numerous degenerated epithelial cells.

The osteopathic changes have been well described by Mr. Jonathan Hutchinson and Dr. Alexis Thompson, and excellent illustrations of the conditions present in their specimens are given in Bramwell's "Atlas of Clinical Medicine," vol. III.

Both skulls show the over-development of bone to be limited strictly to the territory of the trigeminal.

Mr. Hutchinson describes the condition as one of overgrowth of bone, with broad-based exostoses. The overgrowth involves the frontal and sphenoid, as well as the facial, bones proper.

The hypertrophy in this case was said to have become conspicuous in middle life, but Mr. Hutchinson is of opinion that the case was probably congenital. Dr. Thompson's specimen was obtained from an insane patient who has been the subject of epilepsy since childhood. The overgrowth appeared to have commenced in early life, and not to have been congenital. It affected all the facial bones, and the anterior two-thirds of the cranium on the left side.

The convolutions on the anterior and superior aspect of the frontal lobe on the same side were atrophied by the pressure of a large exostosis projecting from the inner table of the frontal bone. To this Dr. Thompson attributes the

epileptic seizures which occurred during life. A similar condition is described in the autopsy on Friedreich's case already referred to (No. 2), and in the second of the cases reported by Brissaud and Lereboullet (62).

PATHOGENY.

In the absence of pathological fact it is natural that speculation as to pathogeny should be exuberant.

The following theories are among those that have been put forward :—

(1) Vaso-motor.—The overgrowth being the result of vaso-motor paresis throughout the affected part, causing vascular dilatation and consequent hypernutrition and hypertrophy. With this view must be associated the theory of sympathetic irritation which has been favoured by several observers, *e.g.*, Barwell (20) and Hermanides (38).

The vaso-motor theory has the support of many writers, such as Trelat and Monod (5), Ziehl (10), Leblanc (41), and others, who base their opinions on the well-known experiments of Claude Bernard and Schiff (42), and on the observations of Stilling (14). In its favour may be cited the frequent vaso-motor derangements, vascular ectases, cutaneous hyperæmias, alterations in surface temperature, salivations, local sweatings, &c., which have been recorded in connection with the condition.

Both the vaso-motor and the sympathetic theories, however, only put the chief difficulty one step further back, as they do not explain why the vaso-motor paralysis, or sympathetic disturbance, arises.

Moreover, it must be noted that in many cases of facial hypertrophy, *e.g.*, those of Fischer, Dana, Montgomery, and my own, there has been a complete absence of vaso-motor indications of any kind. These considerations suggest that while the vaso-motor mechanism frequently shows signs of being implicated in the disease, it is probable that the implication is a secondary condition only, and that it occurs, when it is present, as the result of some deeper lying primary

disturbance of the central nervous mechanism. A similar relation may be observed in the vaso-motor disturbances which accompany syringomyelia.

(2) The theory put forward by Fischer (9) of circulatory stasis in the affected part, produced by vicious positions of the foetus in utero, and setting up hypernutrition and consequent overgrowth. Fischer supported his view by the observations of Miram (43) on the existence of hypertrophy in the distal parts of limbs in which congenital furrows were present.

This theory is obviously inadequate, inasmuch as it leaves the postnatal cases unexplained.

(3) Lannelongue's (44) view supposed some unknown stimulus or irritation of epiphysial cartilages and osteoplastic periosteum, which led to the overgrowth, first of bony, and secondly of soft, parts. This, too, must be put aside as insufficient to account for the cases in which the soft parts alone are affected.

(4) Messrs. Sabrazés and Cabannes (*loc. cit.*) have put forward a teratological theory, and consider the facial hemihypertrophy, when congenital, to be of the nature of an "anomaly by excess," which takes place in the frontal buds at an early date of intra-uterine life. They consider also that other factors may come into play, *e.g.*, variations in the size of the nutrient artery supplying the part.

This view, however, again fails to account for the "acquired" or postnatal cases, which, as we have already seen, appear to be not essentially separable from the congenital.

(5) The trophoneurotic theory. Lewin (13) appears to have been the first to put forward the clinical evidence in favour of a trophic, as distinct from a vaso-motor, theory, in accounting for local atrophies and hypertrophies.

The conflict which he mentions as existing then (1884), between the physiologists and the clinicians, regarding the existence of trophic influences generally, appears to be still unsettled.

Thus, Dr. Aldren Turner, in discussing the pathology of "Facial Hemiatrophy" (Allbutt's System, vol. vi., p. 488),

dismisses the older views, including the "tropho-neurosis" theory of Romberg as scarcely worthy of consideration, while Mr. Jonathan Hutchinson, in the most recent discussion of the same subject ("Atlas of Illustrations of Clinical Medicine"—Fasciculus xix., 1904), after demonstrating that in a great majority of cases, if not in all, facial hemiatrophy is a late stage of fifth nerve morphœa, concludes that it is almost certain the primary sclerodermic changes are brought about under the influence of the nervous system, and that to them, therefore, the term tropho-neurosis is undoubtedly applicable.

The difference of opinion need not concern us here. Whatever the views of the physiologists may be regarding the conceptions of trophic centres and trophic influences, it is clear that the clinician and the pathologist, with the facts of syringomyelia and acromegaly before their eyes, cannot well do without them.

It may be well before proceeding further to recapitulate briefly the various forms in which hypertrophy of different parts of the body occurs.

As Anderson (45) has remarked, general, *i.e.*, complete bilateral, hypertrophy of the body is, at present, a possibility only. It may be considered that giantism, when free from taint of acromegaly, and when the bodily proportions are symmetrically preserved, would exemplify this class; but I am not aware of any authentic record of giantism which fulfils these conditions.

Complete unilateral hypertrophy, *i.e.*, of one half of the body, has of course been frequently recorded, as also has the partial unilateral type. In the latter form the face may or may not participate, and the hypertrophy may affect any limb, or segment of a limb, as in podo- and cheiro-megaly.

In the crossed variety of hemihypertrophy, various combinations may occur. The face on one side may be affected, with the trunk and limbs on the other, or the face and upper limb on one side, and the lower limb on the other.

The hemifacial variety of hypertrophy is thus seen to be merely a partial type, which may exist alone, or with which may co-exist any other form or degree of bodily overgrowth.

As regards the period of onset, whilst most instances in each class are congenital it is clear that all are not so. In Tilanus's case of complete unilateral hypertrophy the difference in size was not apparent till the child was 3 years old. In MacGregor's case (*Glasgow Medical Journal*, March, 1894) the partial hypertrophy apparently commenced in the leg at the age of 6, and in the hand at 10. In Montgomery's case, and in my own, photographs of the patients taken in early life show no existing facial asymmetry.

It will be noted that only in the hemifacial type does the area of hypertrophy correspond with the distribution area, complete or partial, of a peripheral nerve. In the bodily types, those, that is, affecting the trunk and limbs, the hypertrophied regions coincide with no area of nerve distribution, peripheral or radicular. The distribution of the hypertrophy is in these cases segmental or metameric, the boundary line, being, in the trunk, at right angles to the spine, and in the appendages, at right angles to the long axis of the limb. This apparent anomaly may perhaps be explained, as has already been suggested, by Gegenbauer's statement that the trigeminal, the nerve-area chiefly concerned in facial hemihypertrophy, represents the (unilateral) afferent fibres of the first cranial metamere.

It is in connection with this segmental distribution of the hypertrophies of the trunk and limbs, and, presumably, of the face as well, that Brissaud's theory of spinal and cerebral metamerism in nervous disease is of interest, through its conception of the cerebro-spinal axis as a series of neural metameres, or myelomeres, corresponding to and controlling as regards some nerve functions, *e.g.*, the trophic, the aboriginal metameric segments of the body.

Equally attractive as a theory, if less well founded on embryological fact, is the suggestion of outbuddings from the neural axis, in the brachial and crural regions, by which it is sought to explain (47) the distribution of those dystrophic conditions which affect segments of a limb only.

If now we enquire whether clinical observation warrants the supposition of a trophic influence inducing local overgrowth, and emanating from the central axis, it must be

admitted that the facts of syringomyelia and of acromegaly support such a view.

While the dystrophies observed in connection with syringomyelia are most commonly of the atrophic type, it is well known that hypertrophic conditions have not infrequently been recorded.

Chauffard and Griffon (48) describe the case of a man of 48, who had observed for some years an augmentation in the volume of the right arm. Along with other signs of trophic disturbance (ichthyosis, hypertrichosis) the right upper limb was enlarged from the shoulder to the fingers, the bones being also affected. Dissociation of sensation was present in the whole length of the hypertrophied limb, and, in addition, in the shoulder, the right half of the neck, and right half of the cranium and face, the area of disturbance in the latter being limited accurately by the middle line. Perspiration was diminished over this area. No symptom of acromegaly was present unless the enlargement of the hand be so considered.

Somewhat similar cases have been described by Marie (49), in which, with syringomyelia there was present hypertrophy of the right arm and left foot, and by Charcot and Brissaud (50), in which enlargement of the left hand alone was present. Although designated "pseudo-acromegalic," the acromegalic symptoms in the above cases were limited to the enlargement of one or more extremities. In Lountz's case, however (51), there was hypertrophy of the right arm, including the bones, no suggestion of acromegaly being present, and a similar case is reported by Heldenberg (52) in which a man of 27, the subject of syringomyelia, developed hypertrophy of the right arm after an attack of acute rheumatism.

These cases show that there is sufficient clinical warrant for asserting an association between some forms of partial hypertrophy of the body, and lesions of the central gray matter of the neural axis.

The absence of hypertrophy of the cranial and facial bones in Chauffard and Griffon's case was remarked on by these authors, seeing that the dissociation symptoms extended to the cranium and face.

FIG. 5.

Radiogram of cranium of acromegalic giant, Charles F., to show "ressaut post-lambdoidien." Slight facial hemihypertrophy was also present in this case.

Hypertrophy of cranial bones, however, in cases of syringomyelia where no acromegaly was present, has been reported by Colman (53), in whose case frontal bossing occurred, by Westphal (54), who reported frontal and parietal thickening, and by Collier (55). In the latter case some enlargement of the parietal bone co-existed with remarkable deformity of the cranium.

It may be recalled that in 6 out of the 29 cases of facial hemihypertrophy here collected, frontal bossing was present, and that in two parietal thickening occurred as well.

FIG. 6.

Acromegalic cranium in the Ecole d'Anthropologie, showing "ressaut post-lambdoidien" (Papillault).

(From "*Nouvelle Iconographie de la Salpêtrière*," vol. xv., p. 581, by kind permission of Messrs. Masson et Cie.)

The possible relationships between syringomyelia and acromegaly is a subject far outside the bounds of this communication, but in connection with the latter disease I wish to draw attention to the occipital boss reported in Case 29 of the present series. There is, so far as I am aware, no previous record of hyperostosis of the occipital bone in connection with facial hemihypertrophy. Its occurrence in this case appears to me to be of interest, as suggesting a possible link of relationship with acromegaly.

F. Schultze long ago called attention to the occasional prominence of the external occipital protuberance in this disease. Papillault later (56) described the so-called "*ressaut post-lambdoidien*," a bony overgrowth situated posterior to the lambdoid suture, the gradual development of which he had studied in the acromegalic giant Charles, and which he had also found present in an acromegalic skull.

FIG. 7.

Tracing of skull in Case 29, taken by a strip of lead a little to left of mesial line, showing frontal, parietal, and occipital bossing ("*ressaut post-lambdoidien*"?) The thin lines and suture markings are diagrammatic.

From a comparison of a radiogram of the cranial bones in Papillault's case, with the condition present in Case 29, I am of opinion that the occipital boss in this case is of the same nature as that described by Papillault (see figs. 5, 6 & 7).

A further illustration of the relationship I have now ventured to suggest exists in Dana's case (No. 18), in which the two conditions of facial hemihypertrophy and acromegaly were present in the same individual. Another instance of the coincidence of these two conditions is recorded by Launois and Roy (64). In their account of the acromegalic

giant, Charles F., these authors remark that facial asymmetry was present, the right half of the face being a little less developed than the left half. The patient himself had noticed that for the two months previously the left cheek projected more prominently than the right.

Dana remarks of his case that it might correctly be described as one of somato-megaly with hemiacromegaly of the head. Having regard to the frequency with which the cranial bones are affected, it would appear that some such term as hemicephalomegaly would express more accurately than facial hemihypertrophy the actual condition present, not only in Dana's case, but in many of the cases described in this paper.

The condition of chronic trophœdema, described by Meige in 1898 (57), is one that cannot be left out of account in future studies of facial and other hypertrophies.

Following Meige and his school, this may be described as a chronic enlargement, affecting, without prejudice to the health, one or more limbs, or segments of these, including it may be, one half of the face; presenting outwardly the appearance of an œdema, but characterised by absence of pitting, of pain, and of colour. Sensory troubles may be present, and in one case at least [Mabille (58)], dissociation of sensation has been observed.

The condition may be hereditary, or familial, congenital, or acquired.

Meige ascribes the condition to hyperplasia of the subcutaneous connective tissue, considers that it is related to the hypertrophic conditions met with in syringomyelia, and that it is due, pathologically, to changes in the trophic centres located in the central gray matter in the neighbourhood of the trophic centres for muscles. The local hyperplasia may not be limited to cellular tissue, that of fatty tissue may also occur. In the case of muscle, hypertrophy may also occur, though as a rule the muscular dystrophy takes the atrophic form.

As I have already suggested, it appears probable that some of the recorded cases of facial hemihypertrophy, those, namely, in which the soft parts alone have been affected, may

have been conditions closely allied to, if not identical with those described as facial trophœdema by Rapin and Hertoghe (*vide supra*).

It is difficult in considering the pathogeny of acquired facial hemihypertrophy to avoid reference to the contrasted condition of acquired facial hemiatrophy.

Mr. Hutchinson's views upon this subject, founded upon Unna's account of the histology of the tissues in a case of scleroderma, have already been alluded to. Mr. Hutchinson is of opinion that the original local hyperplasia of subcutaneous fibrous tissue which is the essential change in the first stage of scleroderma, is of the nature of a tropho-neurosis. The resultant arrest of growth in the lateral half of the face which constitutes facial hemiatrophy is thus to be assigned, if not directly, yet remotely and indirectly, to some disturbance of trophic, or nutritional, control akin to that which appears to underlie the changes in facial hemihypertrophy.

While the view that facial hemiatrophy is really an arrest of growth consequent on antecedent morphœa is doubtless true of all cases in which the atrophic changes have been observed before, or soon after, the cessation of period of adolescence, some doubt of the universal truth of this explanation will arise in connection with those cases in which the facial atrophy has commenced at mature age. Such are Bramwell's case of J. D. (59) in which the atrophic signs appeared first in a man of 40, and Hoffmann's case (60), where the first signs appeared in a woman, the subject of inveterate neuralgia, at the age of 42. In these cases the question of arrest of growth can hardly arise. Whether preceded by unobserved sclerodermic changes or not (and none were observed in either case), we have to recognise that at ages such as these there can only be retrogressive changes of formed adult tissues.

If we consider that in cases such as these we have pathologically, the converse condition to that which is present in facial hemihypertrophy, we may go further, and find a close parallel to the condition of acquired unilateral hypertrophy in the three remarkable cases described by Raymond and Siccard (61).

In two of these, concerning a brother and sister aged 30, and 35, respectively, progressive atrophy of one side of the body had been observed, extending from below upward, and involving in succession the leg, thigh, abdomen, thorax, upper extremity, shoulder, and one half of the face, the general health meanwhile remaining unaffected.

It is difficult to avoid in these cases, the conclusion arrived at by the authors themselves, that the condition is one of a trophoneurosis consequent on changes in the neural axis.

The following conclusions appear to be warranted by the considerations now put forward: (1) While nothing is known of the pathogeny of facial hemihypertrophy, the most reasonable explanation of the condition is that which associates it with disturbance of hypothetical nutritional or trophic centres, in the periependymal gray matter of the encephalon.

(2) In some of its features facial hemihypertrophy appears to have relationship with syringomyelia, and with acromegaly.

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