face, the forehead and eyes, do not take part in the change, but remain entirely normal. Nevertheless the patient's face is much disfigured, and later on in the disease may have become almost unrecognizable.

While thus quite gradually symptoms have arisen which make the patient a very pitiable object, and which are bound sooner or later to interfere with his position in society, the sad

Fig. 32.—Facial Expression in Progressive Bulbar Paralysis. (After Leyden, Eichhorst.)

truth dawns upon him that even the functions absolutely necessary for the existence of life are failing. Eating, in which up to this time no trouble was experienced, he now finds difficult. It takes a longer time to swallow the food, and in a later stage even mastication becomes impaired. Not only do the movements of the lower jaw become weaker and less energetic, owing to paresis of the muscles of mastication, but, since the powerless tongue is unable to get the food from between the

cheeks and gums into the region of the pharyngeal muscles, the formation of the bolus is impossible. Spoons, fingers, and the like, have to be used instead, or the patient has to hold his head far back to get the food to slide down. Even drinking causes much discomfort, as the liquid may get into the larynx and thus give rise to violent coughing, or may be regurgitated through the nose, either condition being due to weakness of the pharyngeal and laryngeal muscles.

The implication of the larynx is very distressing, and may indeed become dangerous. The voice at times fails, speech becomes irksome, and the tone is monotonous; production of the higher notes as in singing becomes impossible; later on a marked hoarseness and finally aphonia follow, so that the patient can only express himself in whispers, which, owing to the above-described motor changes, are quite unintelligible. At the same time the absence of a firm closure of the glottis, and therefore the inability to cough forcibly, gives rise to various disturbances in the respiratory apparatus, owing to the disability to dislodge mucoid masses which may have collected in the air passages.

Another symptom which, though not constant, is frequently met with, is the marked increase in the secretion of the saliva. This occurs usually rather early in the disease, and not infrequently such patients are seen going around constantly holding their handkerchiefs to the mouth to prevent the saliva from trickling away. On examination, the secretion is found to be viscid. This flow of saliva is due to an actual increase in the amount secreted, as several careful investigators have shown, though they do not agree as to the exact amount.

Two, three, even five, years may pass before any new symptoms are added to those just described. These, however, progressively gain in intensity, and it is especially the change in the features which becomes more accentuated, owing to the constantly increasing atrophy in the muscles of the lips and the cheeks; the palatal reflexes become markedly decreased and finally lost; the tongue, shrunken and distinctly smaller, lies immobile on the floor of the mouth, and can neither be protruded nor moved in any direction. Fibrillary tremor is then not uncommonly marked. On the electrical examination (which is, by the way, very hard to make), we may find reaction of degeneration in the lingual as well as in the pharyngeal muscles.

The inability to take food properly is usually the cause of death; the patient pines away, and gradually dies from inanition without having the blissful benefit of a dulled consciousness to guide him insensibly through his tormenting sufferings. Only in occasional instances disease of the respiratory organs, caused by aspiration of food, hastens the termination (aspiration pneumonia).

Pathological Anatomy.—There is hardly another disease of the nervous system with the anatomical basis of which we are better acquainted than bulbar paralysis. Duchenne pronounced the process to be a primary pigmentary degeneration, and

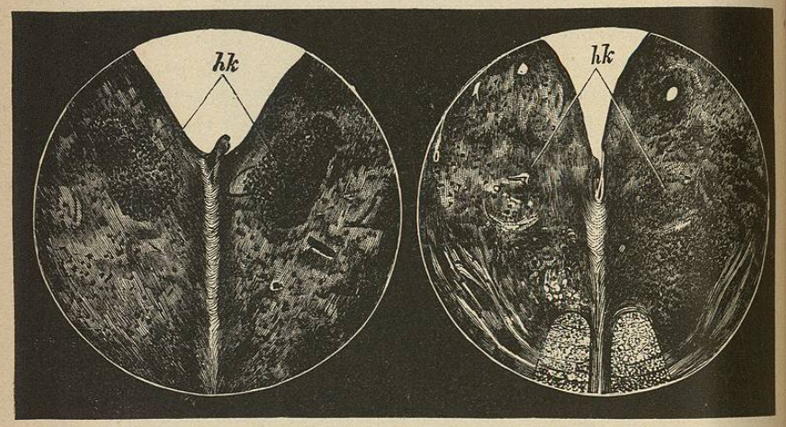

Fig. 33.—Cross-section through the Upper Portion of the Medulla Oblongata. On the left the healthy, on the right the diseased medulla. *hk* on the left the normal, *hk* on the right the diseased hypoglossus nucleus (the nerve cells are almost entirely absent on the right side).

atrophy of the large nerve cells in the nuclei of the medulla oblongata, an assertion which has received complete confirmation from all subsequent investigators. Microscopical examination shows atrophy of the nerve cells. This is shown in Fig. 33 in the nucleus of the hypoglossal; the cells have in this case completely disappeared, having previously diminished in size and lost their processes. At the same time we find the connective tissue increased, the walls of the vessels in the nucleus thickened. Similar changes are found in the nucleus of the vagus accessory and the glosso-pharyngeal nerves (the so-called lateral mixed system, cf. page 107). The former may become diseased in consequence of an ascending

neuritis; a myelitis may be caused by a similar process in the nerves of the lower extremities (Cupfer, Revue de méd., 1890). Since, as we have said before, the upper part of the face always remains normal during the disease, we have to assume that the fibres innervating these muscles arise from a special centre. This is supposed to be a part of the abductor nucleus (Meynert), which has therefore come to be designated by the composite name of facial-abducens nucleus. This and the remaining nuclei, with the exception of those mentioned above, were always found to be intact. The atrophy also extends to the root fibres, which to the naked eye often appear smaller and of a grayish color. From the topographical position of the nuclei below the floor of the fourth ventricle, as it is approximately represented in Fig. 34, we can easily understand how, on the one hand, the pathological process, after having attacked the hypoglossus, next implicates the neighboring vagus, and, on the other hand, how the motor part of the trigeminus usually remains unaffected, so that paralysis of the muscles of mastication is very rare. But why the auditory is constantly exempt and the facial partially affected are circumstances which need to be further investigated. A complete counterpart to bulbar paralysis is found in the so-called progressive muscular atrophy, a disease in which, as we shall see later on, the gray anterior horns of the spinal cord and their nerve cells are affected precisely in the same way as the bulbar nuclei in the disease we are now discussing. The nerve cells of the anterior horns constitute the trophokinetic centres for the muscles supplied by the spinal nerves, an

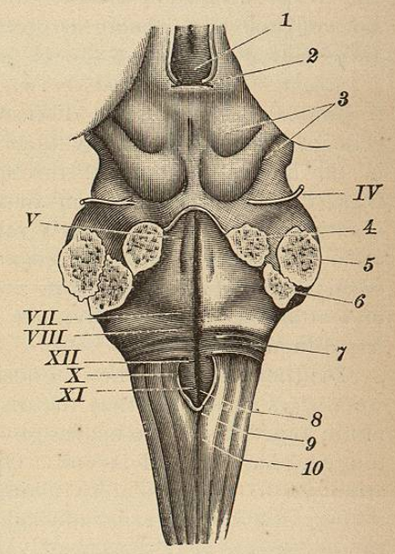

Fig. 34.—The Posterior (Dorsal) Aspect of the Medulla Oblongata. 1, posterior commissure. 2, peduncle of pineal gland. 3, corpora quadrigemina. 4, superior peduncle. 5, middle peduncle. 6, inferior peduncle of cerebellum. 7, striæ acusticæ. 8, funic. teres. 9, obex. 10, funicul. gracil.

office which the bulbar nuclei fulfill for those supplied by the cranial nerves. In both diseases there are atrophy and decrease in the functional power, and in both the disturbance is strictly motor, while sensation is absolutely intact. This essential similarity between the two diseases explains why not rarely one is associated with the other—in other words, why they may complicate each other. We may, indeed we frequently do, meet with cases in which bulbar paralysis is accompanied by atrophy of the muscles of the extremities, while, on the other hand, in progressive muscular atrophy, bulbar symptoms, disturbances in deglutition and speech, may be found.

Another analogy exists between bulbar paralysis and amyotrophic lateral sclerosis, a disease in which not only the nerve cells of the anterior gray horns, but also the motor tract in the lateral columns of the spinal cord are affected. All these diseases, viewed from an anatomical standpoint, if not identical, certainly are closely related to each other, and only differ in the position of the lesions; it is therefore advisable to consider and study them from a common point of view, as the understanding of the individual symptoms will thus be much less difficult.

Diagnosis.—As to the diagnosis, we need not be doubtful if we always remember that the disturbances are confined to the motor functions of the nerves governing the muscles of the lips, tongue, pharynx, and larynx. Oppenheim has recently called attention to rhythmical twitchings of the velum palati and of the internal and external muscular tissue of the larynx, which he considers to be of diagnostic value in diseases of the posterior fossa of the skull (Neurol. Centralbl., 1889, 5). If we find any well-marked sensory changes, if the patient complains of pain or paræsthesias and the like, we either have to give up the diagnosis of bulbar paralysis, or we have to search for some complication. The peculiar facial expression, the increased flow of saliva, the tremulous atrophic tongue partially or even completely immobile as it is, the disturbance in speech and deglutition, when taken together are so characteristic that, if intelligently observed and studied, they will make our diagnosis clear.

There is only one case in which we may be doubtful; certain foci of disease in the brain may produce symptoms simulating bulbar paralysis, so much so indeed that the name pseudo-bulbar paralysis has been given to the condition (which later on

will be described more at length); nevertheless, with due carefulness we can avoid a mistake. The most important point to observe in the differential diagnosis is the course of the disease. While in progressive chronic bulbar paralysis this is slow, but always progressive toward the fatal end, in the spurious form remissions may occur, so that for years the patient may be improved, though he finally also succumbs to the disease. Besides this, pseudo-bulbar paralysis is often attended with cerebral symptoms, headache, apoplectiform attacks, etc.

Prognosis.—The prognosis, as we should expect after what has been said, is altogether unfavorable. There is, according to our present knowledge, no cure for the true bulbar paralysis, and one ought to be careful, therefore, not to deceive the family with promises. As soon as the diagnosis is made they ought to be informed of the unfavorable outlook.

Treatment.—The only treatment from which any success may be expected, if begun early, is the systematic use of electricity: faradization and galvanization of the threatened muscles, especially of those of the tongue and pharynx, frequent excitation of the movements of deglutition, according to the method already described, are the only measures which deserve confidence. With the exception of this local treatment, there is nothing that affords even a temporary benefit. I have never seen any lasting effect from hydrotherapy, but still this treatment is very frequently advised just at that stage of the disease when electricity might do some good. Internal remedies are of no avail; the occasional symptomatic use of atropine (½ to 1 milligramme ($^1/_{120}$ to $^1/_{60}$ gr.) daily) to diminish the salivary secretion may be indicated. It scarcely needs to be mentioned that the chief duty of the physician in the later stages of the disease is to pay the most careful attention to the general nutrition of the patient.

Ætiology.—The ætiology is still obscure. It is true that there are patients affected with the disease who, owing to their occupation, have made rather excessive use of the muscles of the lips, tongue, and palate (glass-blowers, musicians). These cases, however, are so rare that it would seem very forced to attribute any ætiological importance to this factor. The same may be said about syphilis, the truth being that, in most cases, the cause is absolutely obscure, and all we can say is that males and persons advanced in life seem to be more frequently attacked by the disease than others. Heredity but rarely plays

a part, and the influence of cold remains, in connection with this disease, as obscure as with all other nervous affections.

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PART III.

DISEASES OF THE BRAIN PROPER.

THE more autopsies we see the more the fact is brought home to us that brain lesions are frequently present which were not diagnosticated during life. This is by no means necessarily the fault of the diagnostician, for undoubtedly many focal lesions of the brain may exist without giving rise to any symptoms. Recently G. Schmid has published an interesting collection of such cases (Virchow's Archiv, 1893, cxxxiv, 1). On the other hand, of course, we frequently see cases presenting symptoms which make us at once suspect the existence of a brain lesion.

In such cases we have to ask ourselves two questions: (1) Where is the seat of the lesion? (2) What is its pathological nature? To the physician both of these questions are of interest; to the patient, more especially the latter.

The examination which searches for the seat of the lesion will give us the topical diagnosis (τοπος = place); the examination concerning the nature of the lesion, the pathological diagnosis.

The endeavor to localize cerebral lesions—that is, to make a topical diagnosis—has only of comparatively late years received attention, and much of the work so far done can not be called more than an attempt, in many cases indeed only a weak one. The celebrated discovery of Broca (1861), that certain disturbances of speech were often found associated with lesions in the third left frontal convolution, the discovery of Fritsch and Hitzig (1870) that stimulation of certain areas of the cortex produces contractions in certain definite groups of muscles on the opposite side of the body—these and various other, pathological, observations, to which reference will be made later, make it most probable, nay, almost certain, that definite parts or areas of the cortex are always connected with certain func-