and furrows which are found in the face greatly disfigure it (Fig. 124). The corresponding half of the tongue becomes small, and often presents gap-like retractions such as we described in hemiatrophy of the tongue (Figs. 29 and 30). Among the muscles not only those supplied by the facial, but also those

Fig. 124.—HEMIATROPHIA FACIALIS (personal observation).

supplied by the trigeminus, are implicated. The extension of the atrophic process to the neighboring shoulder and even to the upper arm is not unusual. Sensibility is not altered in the affected region.

Most of the few cases observed clinically have never come to autopsy. Of great interest, therefore, was the result of an examination which Mendel was enabled to relate before the Berlin Medical Society, on such a patient who had died of phthisis, and who had previously been examined by Romberg, and later on by Virchow (Deutsche Med.-Ztg., 1888, xxxiii, p. 407). On examining the origin of the trigeminus, all the other roots were found to be normal; only in the descending root could atrophic changes be recognized, a fact which would indicate that the trophic fibres must be contained therein.

Other observations (Ruhemann) also point to a very intimate connection between facial atrophy and the trigeminus; however, more post-mortem observations confirmatory of those of Mendel are needed to clear up the pathology of the disease.

Of the ætiology little that is positive is known. Age and sex seem to have little influence. The disease has been known to occur at all times of life and also to be congenital; it has been observed in both sexes. According to Lewin, the frequency with which the two sexes are attacked is in the ratio of six males to nine females. Hereditary predisposition is certainly not a *conditio sine quâ non*, because there are cases—among others, the one I have reported myself—in which the patients belonged to quite healthy families. Sometimes it has happened that the atrophy was preceded by other nervous affections—trigeminal neuralgia, migraine, epilepsy, etc.—but this is by no means the rule. In a few cases the disease has followed injuries about the face or of the cervical sympathetic (Seeligmüller); more frequently no cause whatever could be demonstrated, and it was impossible to make any conjecture as to the ætiology. The outlook for recovery is absolutely bad, and therapeutics, so far as our knowledge goes, is powerless.

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APPENDIX.—DISEASES OF THE MUSCLES—PRIMARY MYOPATHIES.

Diseases of the muscles, which consist in alterations in their volume (which is generally diminished, less frequently

increased), and consequent disturbances of function, may occur under the most varied conditions. They may be produced by cerebral affections, as we have pointed out on page 232, where we spoke of the possibility of an affection of trophic centres in the cortex; they may be the result of spinal diseases, as we shall later find out in discussing syringomyelia and progressive muscular atrophy, but they may also—and this is what interests us here more especially—occur independently of any central disease as primary myopathies.

Considered from an anatomical standpoint, this disturbance in the nutrition of the muscles, the dystrophia muscularis progressiva of Erb, the myopathia progressiva primitiva of Charcot, consists either in a diminution, a wasting of the muscular tissue, owing to which the volume of the part affected becomes smaller; or during the pathological process there may come about an increase in volume, which is either due to an actual increase and abnormal growth of the muscles—a genuine muscular hypertrophy—or to a growth of the interstitial fatty tissue, in which latter case we speak of a pseudo-hypertrophy of the muscles. Sometimes both conditions are found in one and the same individual, so that certain muscles appear atrophied, while others, in consequence of the simultaneous development of fat, appear strikingly large and hypertrophied. The microscopical examination (Oppenheim and Siemerling, Münzer, and others) shows, besides an increase of connective tissue, a moderate development of fat, and in the pseudo-hypertrophic tissue a considerable increase in the interstitial connective tissue between the individual fibres, which latter have retained their transverse striation (Charcot, F. Schultze, Strümpell). The genuine hypertrophy which is seen in places must be regarded, according to Strümpell, as compensatory.

About the ætiology of primary myopathies we know very little. It should, however, be mentioned that, according to all the observations made up to the present time, they belong entirely to early life, developing as they do before the twentieth year. Heredity plays frequently a *rôle* in the disease, since not uncommonly several cases occur in the same family. Dähnhardt raises the question (Neurol. Centralblatt, 1890, 22) whether there might not occur a lesion of the spinal cord during fœtal life or during the act of birth; if this should be shown to be true, the mother or, as the case may be, parturition will have to be regarded as an ætiological factor.

Lesage has shown that they also may follow certain other diseases, as, for example, typhoid fever (cf. lit.). In such instances we have to deal with a secondary lipomatosis, developing in circumscribed areas of the body, as the result of certain arterial changes.

In our present state of knowledge we seem justified in assuming that these myopathies occur regularly in certain groups of muscles, so that different "types" can be distinguished, and that on the whole the upper half of the body, particularly the

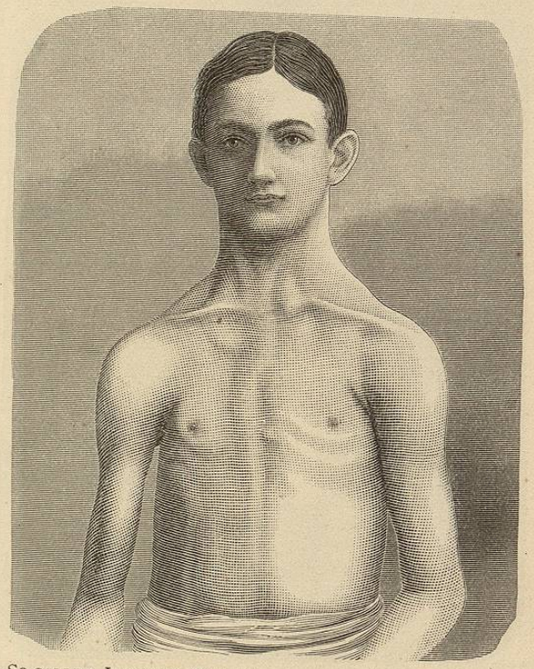

Fig. 125.—So-called Juvenile Muscular Atrophy (Erb) (personal observation).

upper extremities, are more often and more severely attacked by the pathological process than the lower parts, especially the legs. The latter, however, may also be affected, in which case the muscles supplied by the peroneus are especially apt to suffer (Sachs, The Peroneal Form of Leg Type of Progressive Muscular Atrophy, Brain, 1890). It is important always to observe whether the face remains intact or not, as in the former case we are dealing with the hereditary muscular atrophy

which Erb has described as the "juvenile form"; in the latter, with the form which Landouzy and Dejerine have described, and which has by them been called "myopathie atrophique progressive."

The so-called "juvenile muscular atrophy" which develops in early youth, more often in boys than in girls, attacks by

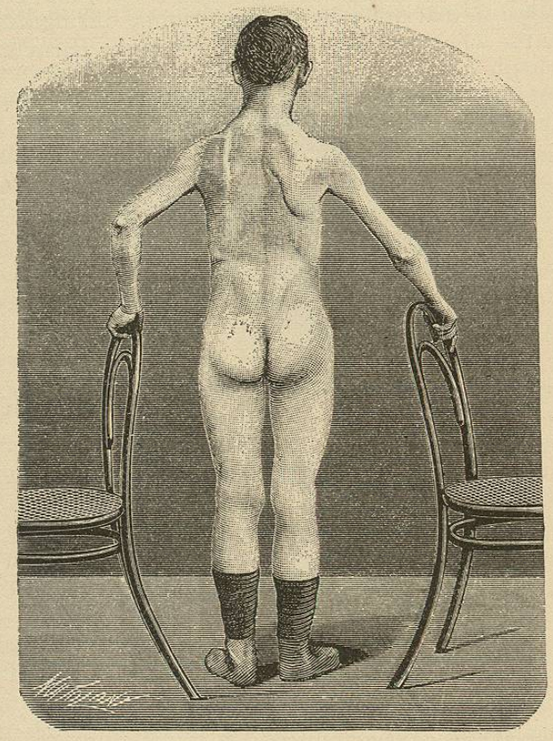

Fig. 126.—Juvenile Muscular Atrophy (Erb) (personal observation).

preference the pectorales, the trapezius, the latissimus dorsi, the serratus magnus, the rhomboidei, the sacro-lumbalis, and the longissimus dorsi, while the majority of the muscles of the forearm, the sterno-cleido-mastoideus, the levator anguli scapulæ, the coraco-brachialis, the teretes, the deltoid, the supraspinatus and infraspinatus, remain, as a rule, intact. The small muscles of the hand, which in spinal atrophy become affected so early and in such a typical manner, are here not implicated

(Fig. 126). It is hardly necessary to enter into a description of the disturbances of function which necessarily must result from disease of so many muscles. If we remember how much impaired are the movements of the arm, which can not be raised above the horizontal position, etc., we can understand the gravity of the child's affliction. If, as in the long course of

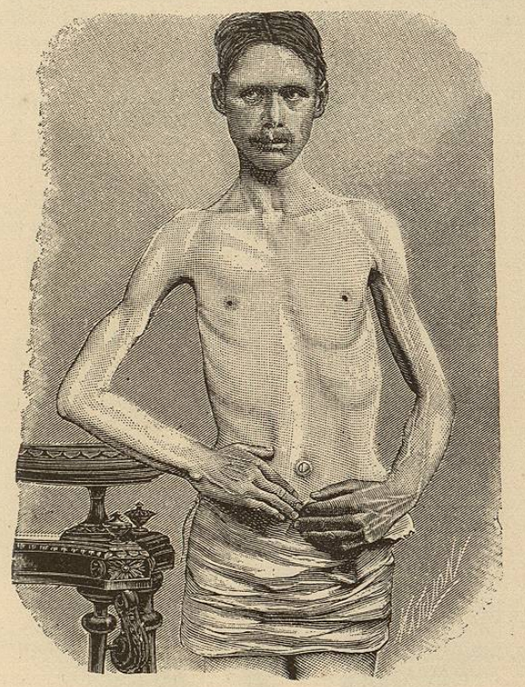

Fig. 127.—Juvenile Muscular Atrophy (Erb) (personal observation).

the disease not uncommonly happens, the process extends to the lower half of the body, the glutei, the peronei, the quadriceps, and tibialis anticus become implicated and the patients at first walk with an uncertain gait, then waddle in a characteristic manner, and finally lose the use of their legs. The implication of the muscles supplied by the bulbar nerves, which has been observed by Bernhardt (cf. lit.), of course has a very decisive influence upon the duration and course of the disease.

Fibrillary twitchings in the affected muscles, as we see it so

commonly, we may say regularly, in the so-called progressive muscular atrophy (spinal), is here wanting with the same regularity. Neither can any changes in the electrical excitability be demonstrated, with the exception, of course, of a diminution due to the disappearance of a more or less large number of muscle fibres. The course of the disease is, as we said, eminently chronic. It may extend over a space of twenty or thirty

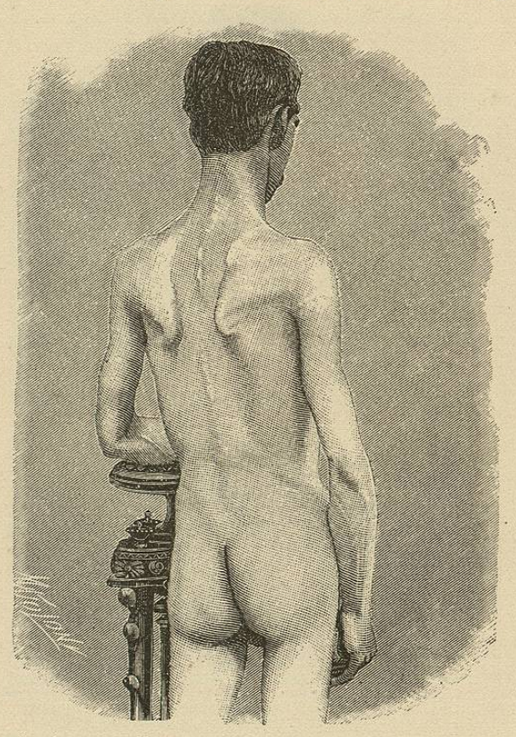

Fig. 128.—Juvenile Muscular Atrophy (Erb) (personal observation).

years, since bulbar symptoms occur but rarely, and we can only look for a fatal issue if the process involves the diaphragm and respiratory disturbances result. The diagnosis never presents any difficulties. After careful examination, taking into account the distribution of the atrophy, the onset of the disease in early life, with the fact that more than one member of the family are affected, the long duration of the disease, the absence of fibrillary twitchings, we can not mistake the myopathic for

the spinal form. The treatment must consist chiefly in good care and nursing. All attempts to arrest the process by the application of electricity or the use of massage, baths, or internal medicines, have proved to be of no avail.

The facio-humero-scapular type of muscular atrophy of Landouzy and Dejerine, the "progressive atrophic myopathy," a form which had, however, already been described by Duchenne under the name of "progressive muscular atrophy of infancy," may manifest itself before the appearance of any other symptoms by a markedly late development of the intelligence (Pillet, Revue de méd., 1890, 5). The atrophy begins

Fig. 129.—Progressive Atrophic Myopathy. Inability to close the eyes completely. (After Marie and Guinon.)

in the muscles of the face, and our attention is attracted by the listless, sleepy expression of the face, the smooth forehead, the faulty movements of the mouth, the inability to whistle and to keep the lips together. At the same time there is a condition of lagophthalmus, so that the patient, in spite of the greatest exertion, is unable to shut his eyes (cf. Fig. 129). Gradually the muscles of the upper extremities and the trunk become affected in almost the same distribution as in the juvenile paralysis. The course is the same in both these forms. Here also there are no fibrillary twitchings and no changes in the electrical excitability, and although the pseudo-hypertrophy of the