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FAMILIAL ULCERS, MUTILATING LESIONS OF THE EXTREMITIES, AND ACRO-OSTEOLYSIS*

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A little over a hundred years ago Nélaton (1852) drew attention to an "unusual disease of the bones of the foot" observed in several members of one family. Similar cases were reported by Smith (1934) and by Cooper, Adair, and Patterson (1947). To the term "familial atrophy of bone" the expression "neurotrophic atrophy" was added. Originated by Duplay and Mort, and later employed by Cassirer, the term merely served to designate a reduction in nutrition of the tissues caused by a peripheral or central nervous disturbance. The later term "trophoneurosis" is even less appropriate, for it implies a functional disturbance of nutrition of nervous origin, although actually referring to severe, irreversible, and even progressive anatomical changes, mainly osteolytic in type.

Another group of cases is characterized by *indolent trophic ulcers.* In the little-known but interesting case of tuberculosis reported by Gastou (1895) in a 51-year-old man whose occupation of window- and façade-cleaner exposed him to cold and injuries, indolent ulcers appeared on the fingers, to be followed by paronychia. A perforating ulcer of the foot and muscular wasting had been present for two years. The diagnosis of syringomyelia was ruled out by the clinical findings and that of leprosy by the results of histological examination. Necropsy revealed tuberculous lesions of the lung, intestine, and meninges, together with what Gastou described as a parenchymatous neuritis occurring in the region of the ulcers. No mention was made of the spinal cord.

Price (1913) seems to have been the first to draw attention to the possible role of *central changes of the spinal cord.* He compared the symptoms observed in his cases with the disease described by Morvan in 1833 as occurring in the fishermen of Brittany. Joffroy, Achard, Thomas, and others have since shown that this disease is marked by the presence of central cavities in the spinal cord, though peripheral neuritis has also been observed by Joffroy and Achard and by Gombault. Thus, Morvan's disease came to be regarded as a form of spinal gliosis associated with peripheral neuritis. Although the author gave the disease a new name, "familial spinal gliosis," and provided it with a new aetiology, he also took into account the possibility of concomitant peripheral changes. A possible familial occurrence was also stressed.

The Syringomyelia Concept

The concept of a familial lumbosacral syringomyelia of the Morvan type based on the similarity of Morvan's "analgesic panaris" with "mutilating trophoneurosis of the feet"—a name recently given to the syndrome under discussion—requires further consideration. The view that syringomyelia is a disturbance of development was first suggested early in the nineteenth century (Calmeil, 1827). However, it was not until after the work of Bielschowsky and Unger (1920), who regarded the disease as a spongioblastosis resulting from incomplete closure of the spinal canal, that the concept of dysraphia developed. The hypothesis was subsequently advanced that a similar developmental disturbance was common to both neurofibromatosis and tuberous sclerosis, and it might even appear as a "microform" in other types of hereditary degeneration (Curtius, 1935). This theory explains the clinical complexity not only of these dysraphic disturbances but also of a whole series of apparently unrelated neurological disorders. Another argument favouring a syringomyelic pathogenesis of this disorder was that osteolysis of the extremities occurs alike in myelodysplasia (as pointed out by Fuchs) and status dysraphicus (Bremer) (quoted by van Bogaert, 1940), both conditions being due to faulty closure of the neural tube.

Several authors, however, maintained an attitude of reserve in regard to such a unitary view. Thus Kienböck (1930) preferred to speak of "a myelodysplastic trophopathy of the foot." He differentiated this condition from syringomyelia, but regarded it as being more or less identical with Fuchs's myelodysplasia.

Clinicians had been particularly impressed by the loss of pain and temperature sense ("syringomyelic dissociation") in the affected extremities at a certain stage of the disease, but their statements became hesitant and less positive as soon as the sensory disturbances detected were found to be more generalized. The whole clinical picture, however, does not correspond very well with classical syringomyelia or with the few genuine cases of familial syringomyelia which have been observed.

Lacking a satisfactory definition, I also classified a case of this type as "lumbosacral syringomyelia" in 1940.

The patient, a member of the De B. family, was affected with symmetrical mutilating lesions of the joints (Fig. 1). A relative had a unilateral perforating ulcer of the foot and trophic ulceration, with a syringomyelic dissociated sensory loss of gauntlet distribution. Another relative had had a perforating ulcer of the foot; the lesion had disappeared, but absence of the ankle-jerk and a similar localized loss of temperature sensibility persisted. Neither of these two relatives had lesions of the metatarsals, but the symptoms observed in this family were far more extensive than those usually seen in myelodysplasia.

Michel André (1941) showed that those clinical features of myelodysplasia which differentiate it from familial perforating ulcer of the foot are principally bony abnormalities due to diaschisis of the spinal cord, the absence of a familial incidence, the early onset of symptoms, and a predisposition to sensory spinal nerve root disturbances of the syringomyelic dissociation type, though the last-named are not normally very severe.

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Hicks (1922) confirmed that familial perforating ulcer of the foot is an independent morbid entity. His patients comprised several members of one family. Hicks was unable to reconcile these symptoms with those of any hereditary or nervous disease or any other disease known to give rise to perforating ulcers. There was some close resemblance with syringomyelia. "The disease is hereditary

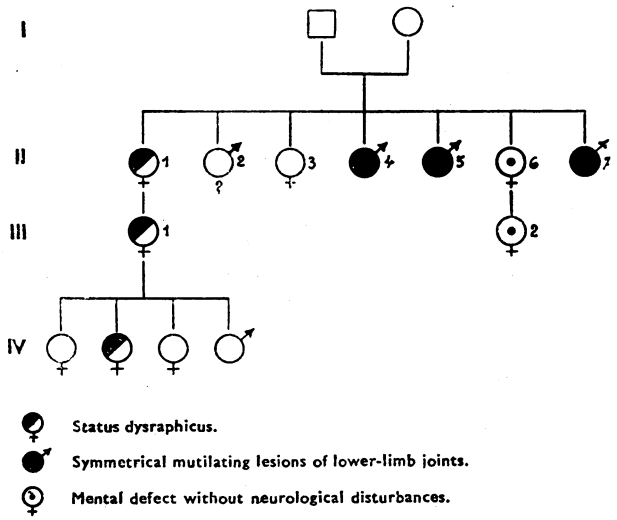

FIG. 1.—Genealogical tree of Family De B. The proband is II 7.

in type. The chief symptoms are perforating ulcers of the feet, fulgurant pains in various parts of the body and deafness."

Thévenard (1942) compared the cases reported in the literature with his own observations. Because of the almost total absence of amyotrophy and motor disturbances and the involvement of the upper and lower extremities, he had strong doubts about the relationship of his syndrome with lumbar syringomyelia. Even to-day the problem is not entirely settled, as there are the familial cases observed by Barraquer-Ferré and Gispert-Cruz (1943) with atypical syringomyelia. In one case the findings at necropsy included a huge cavity in the lumbar enlargement of the spinal cord, the spinal parenchyma being reduced to a thin layer. Unfortunately, the specimen was mislaid and not studied histologically. Again, the case just described (that of the De B. family), in which ulcers and mutilating lesions of the extremities coexisted with a classic familial form of syringomyelia, is not the only one of its type, for identical cases have been reported by Mankowsky and Czerni (1933).

Status Dysraphicus

As signs of status dysraphicus are frequently observed in families or subjects affected with ulcers of the extremities, we shall merely mention the syndrome in passing. There are families, such as those described by Tocantins and Reimann (1939) or Murray Jackson (1949), with numerous dysraphic symptoms. The parents of the generation with "anæsthetic" mutilating lesions of the extremities described by Nagib Taleb (1950) showed symptoms of status dysraphicus and marked inbreeding. The patient affected with mutilating lesions of the extremities in Case I of Velluz, Coirault, and Causse (1955) showed a coccygeal dimple, a shortened frenum of the prepuce, a double urethral meatus, trophoedema of the lower limbs, asymmetry of the breasts, a pigmented hairy mole on the right flank, and pigmented naevi on the back. The mother was affected with trophoedema and perforating ulcer of the foot. Case II of these authors concerned a patient with bilateral Dupuytren's contracture, a coccygeal dimple, congenital absence of the xiphoid process, and pigmented naevi on the back. On the other hand, there are cases in which the symptoms of status dysraphicus are quite few in number.

It may be held that ulcers and mutilating lesions of the extremities, status dysraphicus, syringomyelia, and other similar ectodermal dysplasias are all related, being but dysontogenetic variants. Even though symptoms of status dysraphicus be absent or so mild as to be within the limits of normal variations, detailed anatomical study of verified cases of sarcolumbar syringomyelia may unintentionally provide evidence pointing in this direction. In our case concerning the De B. family, we, like Jughenn, Krücke, and Wadulla (1949), were able to detect in the spinal cord twisted and spherular neurofibromata along the vessels accompanying the ependymal canal and the base of the anterior sulcus, as well as aberrant neuroglial brushes in the posterior columns and hypertrophy involving the collagenous fibres of the pia mater and spinal nerve roots.

It has yet to be established that these "minimal" malformations do not occur in all regions. That they were observed in the spinal cord and its coverings was due to the fact that they are readily detectable in this region, where they are crowded into a small space. The same remark applies to the spinal ganglion. It is extremely difficult to detect similar rudimentary malformations in a sole of the foot which has undergone changes due to suppurating ulcers. This has only been done accidentally when removing material for biopsy. Our pathological investigation has been based on exploratory probing rather than a methodical study of the actual substrate.

Anatomical Data

Before 1949 the delineation of an independent disease was based on clinical data alone, save for the case reported by Gastou (1895), which was not of the familial type, and the valuable but unusual case described by the Barraquers and Gispert-Cruz. The condition was first verified anatomically by Jughenn *et al.* (1949). Their case concerned a 32-year-old woman with indolent ulcers, spontaneous fractures, acrocyanosis associated with vasospasm, camptodactylia of the little finger (as in status dysraphicus), symmetrical ankylosis of the metacarpophalangeal joints of the thumb, malformations of the teeth and palate, numerous pigmented naevi, obesity, and scanty menses. The father showed ulcers and amputation of the toes, bilateral Dupuytren's contracture of the hands, and symptoms of status dysraphicus with syringomyelic dissociation in the lower limbs. One sister presented endocrine disturbances, ulcers, and fractures similar to those seen in the patient. In addition, the two sisters showed marked circulatory disturbances due to paroxysmal acro-asphyxia. A younger sister was affected with diencephalic obesity.

It is because of the circulatory disturbances in the extremities that the paper by Jughenn *et al.* (1949) bears the title: "Contribution to the study of a Familial Syringomyelia: Anatomical and Clinical Studies on a Neurovascular Familial Dystrophy of the Extremities." They say, "Being an important anatomical contribution, what should be stressed to begin with is that syringomyelia may be positively ruled out as a substrate of this syndrome." It is an interesting point that there were slight structural changes, as those in status dysraphicus, as well as gliosis and perivascular neuromas. The latter features also occur in spinal neurofibromatosis. The dorsolateral nuclei of the ventral cornua of the lumbosacral portion of the spinal cord showed tigrolysis, indicating a corresponding peripheral lesion, but the spinal ganglia and the distal portion of the autonomic nervous system were not studied. The peripheral changes involved both the vessels themselves (parietal changes and proliferation of the small arteries to the point of obliteration, thromboses rendered permeable again) and the vasa nervorum; hence the areas of segmental demyelination and proliferation of the white substance of Schwann. The skin and muscles showed a mixture of atrophic and hypertrophic lesions.

The authors concluded that these cases should be classified somewhere between the myelodysplastic and tropho-vasomotor syndromes, possibly along with Kehrer's group of

"hereditary universal acrodystrophies." They believed this term to be questionable, however, as the disease is not merely "acral," but also involves the mid-portion of the foot and the entire distal portion of the segments.

The Peripheral Nervous System

In this way the paper by Jughenn *et al.* focused the problem on the peripheral nervous system—though the lesions of the spinal cord which are evidence of the effects of the disease were not overlooked. Denny-Brown (1951) deserves credit for laying his finger on the primary degeneration of the spinal ganglia and thus broaching a new subject: that of the "polyganglionic and radicular hereditary diseases." The case studied by Denny-Brown and McArdle concerned a member of the family previously described by Hicks (1922). In his anatomical study Denny-Brown (1951) described changes in the spinal ganglia, the posterior columns of the lumbar portion of the spinal cord, the posterior nerve roots, and the peripheral nerves. He also drew attention to an amyloid degeneration of the blood vessels (secondary, in his opinion) and to deposits of some hyaline substance, other than amyloid substance, in the ganglia. Of three cases observed in members of the De B. family, two also showed clinically central deafness, and in the seven years during which we followed up this family after our paper was published in 1939 one of the members developed classical syringomyelia.

In our case we too observed lesions of the ganglia and nerve roots, much less severe lesions of the nerve trunks, and more marked lesions of the peripheral nervous system. The capsules of the ganglia, the perineurium, and the blood vessels showed deposits of a hyaline material staining differently from amyloid substance. Examination of the nerves, skin, muscles, and aponeuroses revealed marked rarefaction of the axis cylinders and fibrosis of the perineurium, subintimal proliferation of the smaller arterioles, and lesions of the muscles of the type caused by neural atrophy. Thus the pathological basis to the condition may extend from an involvement of the nerve trunks, ganglia, and roots, as described by Denny-Brown, to the peripheral nerves, and particular stress might be laid on the effects of these changes on the vascular networks.

So far, the family studied by Hicks and the De B. family have been the only families in which the totality of clinical symptoms described by the English authors was observed. To our knowledge, no other case has since been verified and reported in detail. One of the cases described by Nagib Taleb (1950) is assumed to have been confirmed, but we do not know whether it has been published. The unusual case reported by Girard, Mazare, and Devic (1953) was marked by severe changes of the posterior roots and the trunk of the sciatic nerve, but there were no lesions of the spinal cord, with the possible exception of some slight changes in the posterior columns.

The cases observed by Hicks and by Denny-Brown and McArdle are of value in that they show that there is a hereditary polyganglionic radicular disease giving rise to the clinical picture of ulcers and mutilating lesions of the extremities.

Neuro-ganglio-radicular Changes

The case reported by Denny-Brown and our own case are pure forms of hereditary polyganglionic radicular disease. Should most, if not all, of the familial cases with the lesions under consideration be similarly classified? The problem was stated clearly by Thévenard (1953), who, when discussing pure sensory forms of the disease, wrote: "It may be that this merely is a very early and completely silent stage, but there is no reason for assuming that this sensory dystrophy cannot persist unchanged throughout the entire life of the subjects affected. The problem can only be solved by prolonged observation of these families. . . . As regards the parieto-amyotrophic forms," he added, "cases marked by transitional lesions and suggesting Charcot-Marie atrophy have been reported." He went on to review the cases of Charcot-Marie atrophy associated with perforating ulcers of the foot reported by Oppenheim, Halliday and Whiting, Puussepp, Alajouanine, Sicard and Scherer (quoted by van Bogaert, 1953), and by Denny-Brown and England (1952). To these we can add the recent case of a family described by Barraquer-Ferré and Barraquer-Bordas (1953). In all these families perforating ulcers of the feet appeared in characteristic, if not severe, cases of the peroneal neuromuscular atrophy of Charcot, Marie, and Tooth.

Familial perforating ulcer of the foot may, however, be the first indication of a progressive hereditary degenerative disease of the spinal cord; in other words, the clinical symptoms of the neuro-ganglio-radicular lesion may have advanced to a stage beyond that of all the symptoms of disease of the spinal cord. Thus a painless lesion of obscure origin appeared in the same toe in possibly monozygotic twins at the ages of 10 and 11. *X*-ray examination revealed partial dislocation of the second phalanx and rarefaction of bone. The children had never been able to walk normally and their psychomotor development had been retarded at an early stage of life. At the age of 12, however, they had increasing difficulty in walking and the legs showed incipient signs of becoming thinner, especially on the left side. At the age of 14 the twins showed the picture of Friedreich's ataxia (Fig. 2), accompanied by distal amyotrophy, impairment of temperature and pain sense, loss of deep sensibility, indolent symmetrical ulcers of the feet, trophic disturbances of the nails, subungual excoriations, exfoliative keratosis of the palms, and callosities on the dorsal surfaces of the second interphalangeal joints of the fingers. The clinical picture was that of amyotrophy with severe involvement of the posterior columns. There was an extreme impairment of deep sensibility, involving all four limbs, though the degree of ataxia was not proportional to the severity of these disturbances (Boudin and Djindjian, 1951).

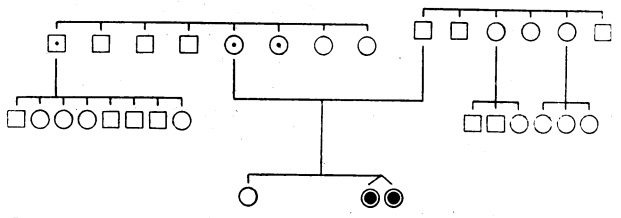

FIG. 2.—Genealogical tree of Family N.

The trophic disturbances in the upper limbs mainly implicated the skin, the hair, and the nails, but minute ulcers were seen to appear on the tips of the fingers.

In these twins, then, the ulcers and mutilating lesions of the extremities appeared prior to the degeneration of the spinal cord, an observation which justifies Thévenard's previous reservations.

These two cases show another characteristic feature—namely, that the disease was associated with oligophrenia, as described by Passouant, Vallat, and Temple (1951).

These findings are important in that they are the converse of those stressed by Denny-Brown and England (1952) and by Barraquer-Ferré and Barraquer-Bordas 1953), another

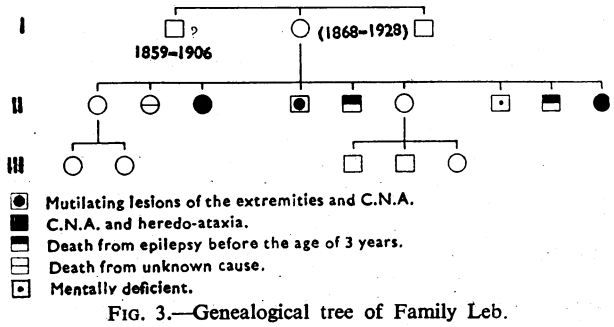

FIG. 3.—Genealogical tree of Family Leb.

instance of which we ourselves have observed. In another family (Leb.) concerning which Troch and Cleen (1953, personal communication) wrote a brief preliminary report, the neuro-ganglio-radicular changes were the last to appear in a long sequence of degenerative lesions of the spinal cord and cerebellum. This Leb. family (Fig. 3), studied since 1922, developed diseases such as heredo-ataxia or Charcot–Marie–Tooth's neuromuscular atrophy (C.N.A.) at various stages of

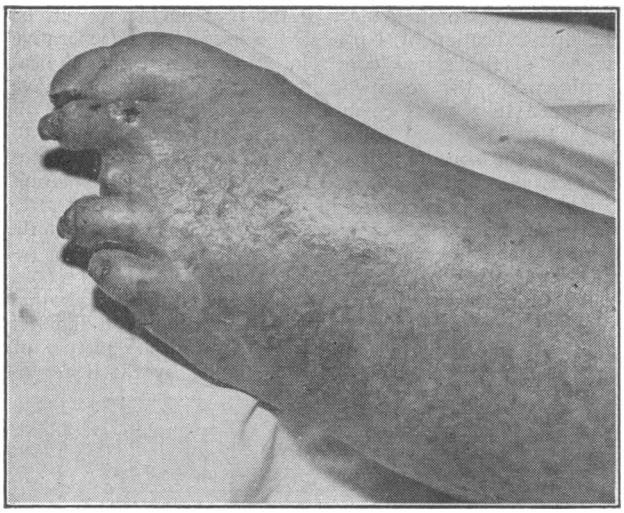

FIG. 4.—Mutilating acropathy of one of the patients from Family Leb.

life, and at the time of writing the eldest son is bedridden, ulcers and mutilating lesions of the extremities (Fig. 4) having been superimposed upon the degeneration of the spinal cord, cerebellum, and nervous system (van Bogaert, 1951).

Related Conditions

Some of the cases in these two clinical groups with ulcers and mutilating lesions of the extremities bear a resemblance to status dysraphicus (including syringomyelia and neurofibromatosis), others to hereditary degeneration of the spinal cord. These conditions appear in young subjects, the average age of onset being 14 (Thévenard, 1953).

With Jughenn *et al.* (1949) we believe it should be stressed that some of these cases are allied to vascular disturbances of the extremities, themselves possibly reflecting a disorder of the peripheral nervous system, and in any case aggravating the course of the disease. We have found no evidence to suggest that *the disease was congenital* in any of our cases; nor did the onset occur during the first two years of life. A recent case (Eyckmans, Radermecker, and van Bogaert, 1956), so far as we know the only case of this type to have been reported in the literature, was characterized by severe mutilating lesions of the extremities, resulting in extensive necrosis of the four extremities at the age of 42. This case is of interest in view of the appearance of a necrotizing lesion at the tip of the nose, and the fact that an ulcer appeared in a little finger when the patient was 2 years old, followed by spontaneous amputation of this finger after sepsis had continued for a few months, ulceration of a toe, going on to spontaneous amputation at the age of 3, etc. In addition to spontaneous mutilating lesions of the nose, extremities, and ears, there were areas of atrophy of the skin with cyanosis, and symmetrical telangiectases in the buttocks and anterior surface of the thigh. In addition to mild lesions due to myositis fibrosa in the extremities, this patient also showed a syringomyelic sensory dissociation of the type observed in ulcers and mutilating lesions of the extremities. We shall not enter into a discussion of the lesions of the skin, a subject on which there has been no agreement (poikiloderma ? Pick–Herxheimer acrodermatitis atrophicans ? cutis marmorata ? special vascular disease of the skin ?). A case of this type compels us to reconsider the problem from the point of view of the peripheral neurovascular disturbances, previously stressed.

Osteolysis

Finally, there are cases in which osteolysis of the extremities proceeds without spontaneous or surgical amputation from suppurating lesions and without any trace of indolent torpid ulcers. In such cases the lesion is confined to a dissolution of the bones, with sheath-like, sausage-like, or lorgnette-like contractions of the fingers and toes. There are no neurological disorders and the trivial sensory disturbances observed may be attributed to thickening of the skin and keratosis or atrophy of the epidermis. The muscular atrophy seen in these cases may be arthrogenic. The patients affected show endocrine disturbances: the skin is delicate in some cases, parchment-like, glossy, scaly, dry, and wrinkled. The hands are soft and pudgy, the joints loose, and the nails small and brittle. Owing to the contraction of the fingers and toes, the skin folds up "like an accordion" over the affected bones. This withered skin of the extremities is accompanied by atrophy of the dental alveoli, and a tendency towards gynaecomastia, kyphosis due to osteoporosis, and changes of the hair and nails. These patients give the impression of having aged prematurely. A certain number of these cases have been collected by Leger and Ducroquet (quoted by Meumier, 1950) and a sporadic typical case was reported by Meumier (1950).

We have previously attempted (van Bogaert, 1953) to classify such patients into certain groups. What Brugsch has described as "acromicria" is undoubtedly related to this disorder, but this is especially so in the cases reported by Harnasch (1950) and Laroche and Hochfeld (1950), the tubero-hypophysial disturbance being particularly marked in these cases. This particular form of familial dystrophy was described by Hozay (1953), who stressed the early inhibition of growth and the non-mutilating osteolysis of the extremities, with malformation of the face.

We have observed this syndrome in a brother and sister. Here, however, the osteolysis of the extremities appeared at such an early stage that miniature extremities were implanted on a body relatively well developed and proportioned (Fig. 5). The undersized portions subsequently under-

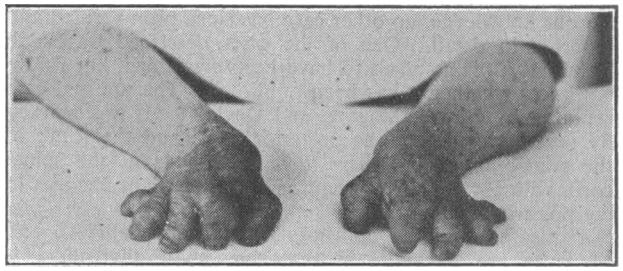

FIG. 5.—View of the back of the brother's hands.

went trophic changes, involving both the bones and the soft tissues. The parents emphasized the fact that the hands and feet of the brother and sister were similar to those of their children up to the age of 3 years and 6 months. At that time the digits became flexed; "not only," they added, "did the fingers stop growing, but subsequently they also decreased in length." The arrest of growth and reduction in size occurred independently: the shrinkage of the bones was not accompanied by the elimination of sequestra through the ulcers. The disease involved the four extremities from the onset. Ulcers did not appear until later, when they showed themselves in friction areas on the soles of the feet, resulting from a faulty position of the twisted stump in the shoes. These traumatic ulcers were painful. They healed within one month. This extraordinary smallness and flaccidity of the hands had its counterpart in a surprising adroitness: the girl could wash up, and the boy was able to roll his cigarettes. They walked about on these

miniature feet, the tiny toes of which continued to be identifiable although reduced to the size of nipples. The reduction in girth and length extended cuff-like as far as the lower third of the forearm, and we are not acquainted with any disease in which arms and forearms as muscular as those of adults are provided with the wrists and hands of a child of 2. The nose and ears had undergone less marked changes in shape. The affected extremities showed no vascular disturbances; the pulsations disappeared on palpation of the arteries. The changes of the skin were a mixture of paper-like atrophy, keratosis, and bands of cyanosis. The nails had been reduced to a thin film and their surfaces corresponded with the size of fingers and toes.

Obviously osteolysis of the extremities had taken on such a grotesque appearance in these patients because it had occurred in infancy. There is no doubt, however, that the other endocrinopathic and trophic changes of the soft tissues elsewhere described and particularly stressed in adults had also occurred in this brother and sister. These endocrine changes, however, are inconstant, for both primary and secondary sex characteristics in the brother and sister described by Hozay (1953) showed a development in keeping with their age.

Changes of the skin, muscles, and vessels as well as the endocrine constitution have not been taken into account in adopting the term acro-osteolysis.

Conclusions

Ulcers and mutilating lesions of the extremities are due to affections of the spinal ganglia, of the spinal nerve roots, and, to a certain extent, of the peripheral nerve trunks and networks. The pathological lesions may predominate in any of these structures. Neurological manifestations are accompanied by changes in the vascular system of variable severity, in the pathogenesis of which several factors probably are operative.

The disease is hereditary in some cases; other forms either bear a resemblance to myelodysplasia in the broad sense of the term or to hereditary degeneration of the spinal cord and cerebellum. The disease may be observed in an isolated state or it may constitute an initial or late stage of hereditary degeneration.

It may be congenital and appear very early in life, be accompanied by extensive and complex lesions of the skin, and be associated with only slight neurological disorders (syringomyelic dissociation of the lower extremities), though of a type similar to that usually observed in young patients.

On the other hand, there are cases of non-mutilating familial acro-osteolysis showing endocrine anomalies, without neurological disturbances, but associated with changes of the soft tissues. Up to now, these cases have not been definitely classified from the clinical point of view.

REFERENCES

André, M. J. (1941). *J. belge Neurol. Psychiat.*, **41**, 340.
Barraquer-Ferré, L., and Barraquer-Bordas, L. (1953). *Acta neurol. psychiat. belg.*, **53**, 55.
—— and Gispert-Cruz, I. de (1943). Quoted by Thévenard (1953).
Bielschowsky, M., and Unger, E. (1920). *Jb. Psychiat. Neurol.*, **25**, 173.
Bogaert, L. van (1940). *Presse méd.*, **48**, 1026.
—— (1951). *Rev. neurol. (Paris)*, **84**, 121.
—— (1953). *Acta neurol. psychiat. belg.*, **53**, 90.
Boudin, G., and Djindjian, R. (1951). *Rev. neurol. (Paris)*, **84**, 252.
Calmeil (1827). Cited by Jughenn *et al.* (1949).
Cassirer, R., and Maas, O. (1910). *Dtsch. Z. Nervenheilk.*, **39**, 324.
Cooper, G., Adair, N., and Patterson, W. M. (1947). *Radiology*, **48**, 509.
Curtius, F. (1935). In *Die organischen und funktionellen Erbkrankheiten des Nervensystem*, edited by F. Enke. Stuttgart.
Denny-Brown, D. (1951). *J. Neurol. Neurosurg. Psychiat.*, **14**, 237.
—— and England, A. C. (1952). *A.M.A. Arch. Neurol. Psychiat.*, **67**, 1.
Eyckmans, R., Radermecker, M. A., and Bogaert, L. van (1956). *Rev. neurol. (Paris)*, **95**, 37.
Gastou, P. (1895). *Bull. Soc. franç. Derm. Syph.*, **6**, 138.
Girard, P. F., Mazare, Y., and Devic, M. (1953). *Acta neurol. psychiat. belg.*, **53**, 82.
Harnasch, H. (1950). *Fortschr. Röntgenstr.*, **72**, 352.
Hicks, E. P. (1922). *Lancet*, **1**, 319.
Hozay, J. (1953). *Rev. neurol. (Paris)*, **89**, 245.
Jackson, M. A. (1949). *Med. J. Aust.*, **1**, 433.
Jughenn, H., Krücke, W., and Wadulla, H. (1949). *Arch. Psychiat. Nervenkr.*, **182**, 153.
Kienböck, R. (1930). *Fortsch. Röntgenstr.*, **42**, 567.
Laroche, G., and Hochfeld, M. (1950). *Sem. Hôp. Paris*, **26**, 1500.
Mankowsky, B. N., and Czerni, L. I. (1933). *Z. ges. Neurol. Psychiat.*, **143**, 701.
Meunier, R. (1950). *Bull. Soc. clin. Hôp. Charleroi*, **1**, 151.
Nélaton, A. (1852). Cited by Thévenard (1953).
Passouant, P., Vallat, G., and Temple, J. (1951). *Rev. neurol. (Paris)*, **84**, 248.
Price, G. E. (1913). *Amer. J. med. Sci.*, **146**, 386.
Smith, E. M. (1934). *J. Amer. med. Ass.*, **102**, 593.
Taleb, N. (1950). *L'acropathie ulcérante familiale.* Faculté française de Beyrouth (thesis).
Thévenard, A. (1942). *Rev. neurol. (Paris)*, **74**, 193.
—— (1953). *Acta neurol. psychiat. belg.*, **53**, 1.
Tocantins, L. M., and Reimann, H. A. (1939). *J. Amer. med. Ass.*, **112**, 2251.
Velluz, J., Coirault R., and Causse, R. (1955). *Ann. Méd.*, **56**, 198.

PERIPHERAL ARTERY GRAFTING

DESCRIPTION OF AN OPERATION

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During recent years there has been increasing experience of the surgery of the aorta and great vessels. Reconstruction of these with a variety of different substances has achieved considerable success, but there is not yet unanimity of opinion on the best material to use. In general an end-to-end anastomosis is used in these vessels. That so many different materials have proved successful is probably due to the large diameter of the anastomosis and the rapid rate of blood flow. In the peripheral vessels, where the diameter of an end-to-end anastomosis is smaller and the blood flow slower, thrombosis is common. Many consider that there is only a limited use for peripheral grafting, particularly when atherosclerosis is the cause of the condition requiring treatment (Fontaine *et al.*, 1952; Rob and Eastcott, 1953). Since the introduction of the end-to-side technique of anastomosis, first suggested by Kunlin *et al.* (1951) and later modified by Cockett (Linton and Menendez, 1955), the outlook in peripheral grafting has improved.

During the past 10 years we have inserted 78 peripheral grafts in the lower extremities, most of them for the treatment of atherosclerosis obliterans, and on the basis of this experience we have drawn certain conclusions.

Materials Used for Peripheral Arterial Reconstruction

Synthetic Materials.—Although many synthetic substances are available, the only material that we have used for peripheral arterial replacement is polyvinyl alcohol sponge. This substance has been used on six occasions; all the prostheses functioned initially, but only one, an end-to-side anastomosis between the aorta and the common femoral artery, remains patent. In our experience polyvinyl alcohol prostheses, though successful in the aorta and iliac vessels,