ON CHRONIC ATROPHIC SPINAL PARALYSIS IN THE CHILD; AND ON A RARE MODIFICATION OF THE REACTION OF DEGENERATION.

BY W. ERB, M.D.,

Professor of Medicine to the University of Heidelberg.

It is not very long since clinical, experimental, and electro-diagnostic researches have made it possible to define and establish the morbid type called "Atrophic paralysis." We understand thereby all such motor paralysis with which is associated a rapid disappearance of the muscles, due to the degenerative atrophy of the motor nerves and of the muscular tissue. The presence of this atrophy is demonstrable by an electrical investigation of the tissues which then present the reactions of degeneration¹ more or less fully developed. Notwithstanding the general uniformity of the picture presented by atrophic paralysis, a closer investigation has shown that it may arise from morbid processes of different nature and variable localisation. Pure cerebral paralysis, however, arising from lesions situated above the pons, are never accompanied with degenerative atrophy and R.D.—(except in the case of certain rare complications and of secondary changes). Degenerative reactions, on the other hand, are frequently observed in numerous kinds of spinal and peripheral lesions, under definite circumstances. Hence, atrophic peripheral paralyses have with right been brought by the side of spinal paralyses.

Every possible lesion of the peripheral motor nerves—provided they be deep enough to create a complete break of con-

¹ [The abbreviation R.D. may conveniently be adopted to stand for "Reaction of Degeneration"—a clumsy expression for which, however, it is difficult to find a substitute at once short and correct.—A. DE W.]

ductivities—may lead to atrophic paralysis. Thus the so-called “rheumatic” affection of certain nerves (e.g. of the facial), but much more frequently the mechanical and traumatic lesions of a nerve (compression, crushing, dividing, tearing), and finally a sufficiently developed neuritis, lead to a complete destruction of the conducting paths in the nerve. The last-mentioned cause, neuritis, has lately been made to assume a much too prominent position; though it certainly occurs in connection with atrophic paralysis both isolated in single trunks, or diffused as “multiple neuritis,” yet its occurrence is not so frequent as some authors believe. It is by no means proved that all which now goes under the name of neuritis is so in reality; post-mortem appearances have often displayed the closest analogy with the degenerative atrophy of the nerve (of neurotic origin), and cannot without further proof be admitted as “neuritis.” It is conceivable that this degenerative atrophy is set up in the nerves themselves (e.g. by certain poisons, lead, ergotine), but it is perfectly admissible also that it depends upon a functional disturbance in the anterior spinal cornua, the nature of which as yet escapes our means of microscopical investigation. Scheube¹ and Bälz,² basing themselves upon good observations and sound reasoning, have advanced the view that the Japanese *Kak-ke*—that peculiar endemic disease which occurs so frequently in Eastern Asia, as well as in other countries (under the name of *Beri-beri*)—is nothing but an infectious inflammation of the peripheral nerves; should this theory be confirmed, multiple neuritis would acquire a meaning of considerable importance. As far as Europe is concerned, however, this fact would perhaps only serve to stimulate thought and research with reference to the few scattered cases of multiple neuritis observed here.

The spinal atrophic paralyses are doubtlessly to be referred, in almost every instance, to disease of the anterior grey matter of the cord—the possibility being excepted, in some rare instances, of a lesion of the anterior roots within the cord,

¹ Scheube, “Die japanische *Kak-ke* (*Beri-beri*),” *Deutsch. Arch. f. Klin. Medicin*, xxxi. and xxxii., 1882.

² Bälz, “Ueber *Kak-ke* (*Panneuritis endemica*),” *Zeitschr. f. Klin. Medicin*, iv. 1882. *Mittheilgn. d. deutsch. Gesellsch. f. Natur- u. Völkerkunde Ostasiens*, part 27, 1882.

before they reach the grey matter. It is still doubtful whether degenerative atrophy of spinal origin implies always destructive lesions, recognisable microscopically by the destruction or degeneration of the large multipolar ganglionic cells. It is, as just mentioned, quite possible that functional, microscopically unrecognisable disturbances of these cells (e.g. toxic substances) may have the same effect. This effect would obviously manifest itself by symptoms of paralysis and rapidly progressing atrophy, with the associated phenomena of R.D.; without implication (in pure cases) of sensibility, micturition, and skin-nutrition; but usually with diminution of the reflexes,—of the tendon-reflexes more particularly.

The chief forms of disease of the anterior horns are the following. Acute atrophic spinal paralysis (*poliomyelitis anterior acuta*), often named spinal infantile paralysis, is a type of disease which calls for no remarks here, having been well characterised in its development, its course, its localisation, its sequelæ, and by a sufficient number of post-mortem examinations with uniform results.

The typical progressive muscular atrophy and its closely allied amyotrophic lateral sclerosis are likewise characterised clinically by atrophic paralysis with R.D., anatomically by destruction of the anterior horns—the evidence being so clear as to leave no opening for any serious objection against their being set up as special forms of disease. It is clear, however, that the question may still be raised as to whether they consist in a true inflammation, or not rather in a slowly progressive primary degeneration; the future must decide upon this point. The form of atrophic paralysis which has hitherto been least investigated is the one known as *Poliomyelitis anterior chronica* (*Paralysie spinale antérieure subaiguë*, Duchenne) and which has been distinguished and defined on clinical grounds chiefly. The attempt made by Leyden¹ to reduce the spinal and peripheral atrophic paralyses to one group, and attribute to multiple neuritis the main part in the production of what has hitherto been understood as subacute or chronic poliomyelitis, does not appear to me very happy. Its result can

¹ E. Leyden, "Ueber Poliomyelitis und Neuritis," *Zeitsch. für Klin. Medic.* i. 387, 1880.

only be to obscure a subject which was already undergoing the process of sifting and enlightening. Leyden falls into the very error which he censures in the supporters of poliomyelitis; a fact novel, or at least insufficiently studied, is thereby prematurely generalised and typified. It is certain that subacute or chronic multiple neuritis exists; and that many a case which has been described as poliomyelitic really belongs to it; but on the other hand, the results of post-mortem examinations¹ leave no doubt as to the existence of chronic poliomyelitis,—as a view strongly expressed by Eisenlohr (loc. cit.) and Remak.² Clinically, the two diseases may usually, in my opinion, be clearly distinguished, and if I may draw a conclusion from my own experience, the poliomyelitic type of disease is of far more frequent occurrence than is multiple neuritis.

Chronic poliomyelitis consists in a more or less creeping and slow development of a paresis or paralysis with atrophy and R.D.; a paralysis which usually has a character of gradual ascent, without any disturbance of sensation beyond slight dysæsthesiæ, or paræsthesiæ, and moderate pains. These phenomena cannot be elicited by an objective examination, and are not accurately limited to the area of distribution of single nerves, but invade more diffusely a whole extremity.

The distinction between chronic and acute anterior poliomyelitis rests mainly upon the slow development of the paralysis,

¹ Apart from the older results, see *Erb*, *Krankh. d. Rückenmarks*, 2nd ed., p. 722, 1883.

Aufrecht, "Ein Fall von subacut. Spinalparalyse," *Deutsch. Arch. f. Klin. Medic.* xxii. p. 33, 1878.

Bäumler, "Fall von Poliomyelit. ant. subac." *Tagebl. v. 52 Versamml. d. Naturf. u. Aerzte in Baden*, 269, 1879.

Edes. "A case of Anterior Spinal Paralysis with formation of vacuoles, etc.," *Bost. Med. Surgic. Journ.* 1879, July 24.

J. Ross, 'Diseases of the Nerv. System,' vol. ii. p. 139, 1881.

Jr. Neumann, "Poliomyelit. anter. subacuta, complicirt mit Leptomeningitis chron.," *Deutsch. Arch. f. Klin. Med.* xxviii. p. 589, 1881.

R. Schulz u. Fr. Schultze, "Z. Lehre von der acut. aufsteig. Paralyse," *Arch. f. Psych. u. Nerv.* xii. p. 457, 1881.

Landouzy et Déjerine, "Des Paralysies générales spinales à marche rapide et curable," *Revue de Méd.* 1882, Août.

C. Eisenlohr, "Poliomyelitis anter. subacuta cervical. circumscripta," *Mendel's Neurol. Centralbl.* 1882, No. 18.

² *E. Remak*. Article: "Spinalähmung," in *Eulenburg's Real-Encyclop.* 1882.

its progress in time, and the more favourable prognosis; in the majority of cases the disease (unless it has an ascending sub-acute character) passes into a regressive stage, and a perfect recovery is obtained—a feature almost never present in the acute form, as Duchenne has already pointed out. This chronic form of atrophic paralysis does not seem to have hitherto been observed in children, among whom the acute disease is so prevalent. Recent literature of spinal diseases contains but negative statements with reference to this point, based on the general authority of Duchenne.¹ Thus Eulenburg,² Bristowe,³ Ross,⁴ and myself⁵ have stated that the disease occurs only between the 30th and 50th year. Remak⁶ says plainly that the disease is never met with in children. Ross himself, however, publishes a case,⁷ with autopsy, of the disease in a girl of 15. Hughes Bennett⁸ gives, under the name of poliomyelitis anterior chronica, an observation taken on a child 2 years of age, but which does not appear to me to belong to the category. (Disease beginning during measles; no marked atrophy of the paretic legs; knee-jerk present; electrical examination of legs not recorded.) The case of Ross would thus be that of the youngest individual affected; no certain instance has yet been recorded of the disease attacking a child.

I have lately made an observation on a child of six years old, which is calculated to fill this gap. Though recovery ensued, and thus the diagnosis has not received its confirmation by autopsy, yet it has been so carefully investigated with a special view to the differential diagnosis, that there cannot be any doubt as to the assumption of a chronic anterior myelitis being justified. In addition, the case is sufficiently interesting in itself, owing to the peculiar manifestation of its R.D., to call for an accurate description and commentary.

Clara Schellenberg, 6 years old, comes from a healthy

¹ 'Electrisation localisée,' 3rd ed., p. 479.

² 'Nerven-Krankheiten,' 1878, vol. ii. p. 381.

³ 'Theory and Practice of Medicine,' 3rd ed. p. 958.

⁴ 'Diseases of the Nervous System,' vol. ii. p. 136.

⁵ 'Rückenmarkskrankheiten.' ⁶ *Loc. cit.* p. 11.

⁷ *Loc. cit.* p. 139. ⁸ 'Electrodiagnosis,' p. 121.

family; mother slightly nervous. The child has had measles and hooping-cough, but otherwise has enjoyed uniform good health, and was always able to walk and romp like other children. Never had scarlatina nor diphtheria. Early in July 1822 the parents noticed that her right foot turned a little inwards, and was slightly dragged. About a fortnight afterwards the left foot showed the same symptoms, which very gradually became more marked, until the gait was greatly altered.

There never was any fever nor general disturbance; the disease was developed very slowly and gradually. Never any complaint of pains during the whole time. Micturition stated to have been a little delayed at first; but the function soon became quite natural again, and remained so. On the 22nd of August the child was brought to my policlinique (then in charge of Dr. Günther), and for the first time examined. The child, who was very intelligent, denied absolutely the existence of any pain whatever. No anomalies of the head, trunk and upper extremities. Both legs markedly paretical; the child cannot walk unless half carried, the toes hang down, the foot is put down awkwardly and unsecuredly; the gait is thereby very tottering. Below the knee there is distinct wasting. No perceptible alteration of sensibility; knee-jerk and plantar reflex absent. Functions of bladder and rectum perfectly normal. Electrical investigation shows partial R.D. in the district of both peroneal nerves. Direct galvano-muscular excitations yield sluggish contractions, but KCC is greater than A C C.

The child is galvanised twice a-week (current "stable" to lumbar region, kathodic "labile" galvanisation of nerves and muscles of legs). The gradual invasion of the disease continues. Paralysis and atrophy increase. The child can no longer walk, but no pains are experienced.

On the 23rd of October, 1882, for the first time, I carefully examined the little patient. The child looks weakly, but is spirited and well developed. Her answers are to the point, and perfectly consistent. Head, trunk and arms normal, legs very weak, and locomotion greatly impaired, owing to complete paralysis of the peroneal districts on both sides; complete

paralysis of the right tibial district; almost complete paralysis of the left tibial district (trace of flexion of toes left). Paresis of triceps extensor and flexors of thigh on both sides, slight on the left side, apparent on right. Functions of bladder and rectum normal.

Sensation unimpaired, as shown by repeated testing with reference to touch, pain, and temperature; the point and head of the pin are everywhere clearly distinguished. Tickling produces no impression on the soles, but neither does it on the upper part of the body. The farado-cutaneous sensation test, applied with my electrode, furnishes normal data, with no greater divergences than usually occur in patients of her age. No diminution worth mentioning in the lower as compared with the higher half of the body, nor difference between the two legs, especially in the districts of the paralysed nerves.

Plantar reflex and knee-jerk absent on both sides. The muscles are quite flaccid and soft. The joints passively moveable. The toes, when raised and abandoned to themselves, fall at once and quickly. Both legs are distinctly wasted: circumference of calf: right, 17; left, 18 cm.—of thigh (12 cm. above patella): right, 26·5; left, 27·5 cm. The muscles are not particularly tender to pressure, but their mechanical excitability is increased (especially in the peroneal district); the contractions obtained by their percussion follow a retarded course, and are tonic in character. In the vastus internus similar contractions obtained, but are rapid. The electrical exploration yielded peculiarly interesting results. I shall condense the results of accurate and repeated examinations of the peroneal district especially.

The faradic excitation of the nerves showed that the minimal contractions occurred at the following distance of the secondary from the primary coil.

N. frontal,	right,	206 mm.—left,	210 mm.	
N. accessor,	"	227 "	"	220 "
N. ulnaris,	"	212 "	"	210 "
N. peroneus,	"	215 "	"	203 "
N. pro vast. int.	"	195 "	"	210 "

These numbers (considered along with the body-resistances estimated on the galvanometer) showed that there was no

demonstrable quantitative alteration in the farado-excitability of nerves. The direct faradic exploration of the muscles gave :

Extensors of forearms :	right, 182 ;	left, 182
Flexors of forearm	„ 190	„ 190
Triceps extensor fem.	„ 155	„ 165
(common motor point)		
Vastus internus	„ 170	„ 195
Vastus externus	„ 155	„ 165
Tibialis anticus	„ 163	„ 160
Extens. digit et peron.	„ 155	„ 145
Muscles of calf	„ 145	„ 155

These numbers show a distinct, though not considerable diminution of the direct faradic excitability of the affected muscles.

The galvano-nervous exploration showed that the first K C C. occurred with the following number of cells and galvanometric deflections :

N. ulnar. ;	right, 12- (3°)—	left, 12- (3°)
N. peron.	„ 12 (3°)	„ 12 (3·5°)

Fourteen cells cause in the peroneal nerve very strong K C C, weaker A O C, very weak A C C.

The galvanic exploration of the muscles showed in the vastus internus distinct K C C with 14 cells ; K C C > A C C, contraction not slow in this, nor in the other portions of the triceps extensor.

Marked R. D. in the anterior tibial, extensor digit. long., and peroneal muscles ; contractions very slow, tonic ; ACC > KCC ; no A O C. Latency of excitation very marked ; the contraction occurs late and lasts an unusual time.

In the calf-muscles excitability diminished, but not so tonic and protracted as in the peroneal.

The general result of this examination is, for the peroneal district, partial R. D., which in itself constitutes nothing strange. The remarkable feature about this case is the fact that the contractions had the characteristic protracted course and tonic duration, not only when obtained by direct galvanic excitation, but presented them also in an equally marked manner when obtained by the (galvanic or faradic) excitation of the nerves, and by the direct faradic excitation of the muscles.

This rare phenomenon was observed from the very first

exact observation made, but was submitted repeatedly to various tests, the results of which were shortly as follows:—The galvanic excitation of both peronei with 12–14 cells (the duration of the closure being only momentary) is followed by a lazy, tonic contraction, which passes off gradually,—not so protracted, however, as that due to direct muscular excitation. The contraction obtained by AO is likewise slow and persistent. On increasing the strength of the current, the line of ascent of the contraction increases in slope and height, but that of descent is always gradual (as is well shown by the contrast with the sudden fall of the foot from the state of passive dorsal flexion). Faradic excitation of both peroneal nerves (secondary current and rapid vibrations of the hammer, either A or K of break-current) produces the same abnormal course of contraction even when the circuit is closed for a very short time. Powerful currents cause a powerful and rapid rise of the muscle, but on opening the circuit the relaxation is still characterised by its sluggish course. The primary (extra) current gives rise to the same phenomena. In order to ascertain whether instantaneous excitations had the same effect, single induction shocks (of secondary or primary) were tried, and found to be followed by contractions having the same gradual rise and fall.

The faradic stimulus directly applied to the muscles themselves gives—if somewhat stronger than when applied to the nerves—the characteristically developed contractions. The alterations are much more accentuated than in the contractions elicited by indirect excitation, and are observable whether rapidly interrupted currents or single shocks are used.

With reference to the order of contractions, repeated observations show that nerves react only to KC and AO ($KCC > AOC$; contraction slow), and that muscles react only to KC and AC ($ACC > KCC$; contraction much protracted and slow).

I shall now give a brief summary of the progress of the case, which went on very satisfactorily. On November 3rd, 1882, the first traces of returning motility in the peroneal district were observed, all the other circumstances, including the electrical phenomena, remaining the same.

November 27th.—Amelioration marked. Motion of left foot extensive and forcible; distinct movements on the right side in the peroneal and posterior tibial districts. The child moves about easily. Triceps extensor and flexors of thigh much stronger, on left side especially. Alteration of contractions still very marked on right side, much less so on the left. Circumference of calf: right, 18 cm.; left, 19 cm. Sensation always perfect; no pain.

December 9th.—The left leg is almost normal in its movements; the knee-jerk has reappeared in it, but not on the right side. Right foot moves better. Electrical contractions approximate the normal type on the left side: 12 cells, slow ACC: 16 cells, rapid KCC. On the right side the anomalous responses still persist.

December 31st.—Continuous, progressive improvement. On the left side the condition is almost normal. Muscles fuller, knee-jerk lively; farado- and galvano-nervous and muscular excitations do not call forth delayed contractions. AOC recedes into the background. On the right: motility much improved, especially in the peroneal district. Muscles still flaccid; no knee-jerk. Electro-muscular excitability diminished, but still protracted responses to both direct and indirect excitation.

January 19th, 1883.—Further progress. Left leg quite normal. Right still weak, drags readily; still the child goes about in a lively manner; knee-jerk absent. Electrical phenomena returning to the normal; contractions much less protracted. ACC still prevails slightly in muscles. Faradic excitability of both peroneal nerves equal (203 mm.). Farado-cutaneous exploration gives the same results as formerly. Muscles firmer and fuller. Circumference of calf: right, 18.5 cm.; left, 19.5 cm. Thigh (12 cm. above patella): right, 28 cm.; left, 29 cm. General condition excellent; feet less cold.

Commentary.—The diagnosis of this case—unique, *quoad* the age of the patient—cannot be doubtful in my judgment. We have to deal with an atrophic paralysis of gradual and progressive development, and with partial R.D. in the lower extremities, without the trace of any anaesthesia, paræsthesia, or pain; a paralysis which does not consist in the successive

implication of the nerve-trunks involved, but which invades the whole lower extremity in more diffuse and with diminishing intensity as it ascends upwards. The sphincters are free; the general state is not disturbed. It presents, therefore, the typical image of a spinal atrophic paralysis; a multiple neuritis is excluded by the absence of every sensory disturbance and the diffuse distribution of the paralysis. The most obvious supposition is that of a lesion (inflammatory?) of the anterior horns of the lumbar enlargement, most severe in the lower half of it (nuclei of origin of the sciatic nerve), and diminishing in intensity as it ascended into the upper half (nuclei of the lumbar plexus), the right side being more deeply affected than the left. Such is the disease which we call chronic anterior poliomyelitis. The case before us belongs more particularly to the class of milder cases forming the "middle form" of poliomyelitis previously described by myself;¹ but I should no longer consider it necessary to give a special description of this form, which obviously is but a variety of a single morbid type.

The favourable course of the disease, culminating in complete recovery, is an important characteristic element of it; a course frequently observed among adults, in opposition to the acute type of poliomyelitis, or infantile paralysis, from which it may be diagnosed by the partial R.D. and this favourable course, as well as by the kind and mode of its development.

The result of the galvanic treatment, the only one employed, deserves some remarks. Whether we must ascribe to it the fact that the left leg, which was the last attacked, recovered first, is of course not easy to determine; and this observation cannot be considered as a proof of the value of galvanisation. Still such a possibility deserves to be noted.

Of greater interest still is the peculiar behaviour of the partial R.D. in the nerves and muscles involved. The main point is that every kind of direct and indirect excitation of the muscles, even instantaneous stimuli, invariably produced sluggish contractions. This is usually not the case with partial R.D., such contractions occurring only upon direct galvanic

¹ Erb, "Ueber eine noch nicht beschriebene 'Mittelform' der Chron. Atr. Spinalähmung." *Centribl. f. Nervenheilkunde*, I. 3. 1878.

excitation of the muscle; its faradic excitation, like its indirect excitation by either current through the nerve, yields only the normal rapid, short-lived, contraction. But all this was altered in our case.

These phenomena, however, are by no means absolutely new, they have been previously observed. I have myself in a paper on Peripheral Paralysis¹ described a case of ulnar paralysis in which faradisation of the muscles, and later of the nerves, called forth a distinctly retarded contraction. E. Remak² observed the same phenomena in a case of sub-acute atrophic paralysis, and has given to this sub-species of partial R.D. the name of "Faradic R.D."

Two of my former assistants³ have related cases of neuritis which occurred under my own observation in which this peculiarity occurred. In one of the cases the contractions were lazy when obtained by galvanic excitation of the nerves.

Kast⁴ has also described quite lately a case in which a certain diagnosis (peripheral or poliomyelitic lesion) could not be arrived at, but where the same anomaly was found. The contractions obtained by faradisation of nerve or muscle were protracted. The galvanic excitation of the nerve, however, appears to have been followed by rapid and short-lived contractions.

Finally, I have observed the phenomena in several cases of paralysis of various origin ("rheumatic," in facial; traumatic, in median; pressure, in musculo-spiral); it was more or less accentuated, occasionally mere traces of it were present. It would thus appear not to be so very rare, and I am inclined to think that careful investigation will prove it to be less rare still, especially as a temporary or transitional condition in the course of the disease.

With reference to the question as to the meaning of this peculiarity in the partial R.D., I can but subscribe to the

¹ Deutsch. Archiv f. Klin. Med. 1868, iv. pp. 554, 556. See also my 'Elektrotherapie,' p. 210.

² Arch. f. Psych. u. Nervenkrankheiten, 1879, ix. p. 551.

³ Kast, "Zur Lehre von der Neuritis." Arch. f. Psych. 1881, xii. p. 266. Vierordt, "Ueber atr. Lähmungen der ob. Extremitäten." Deutsch. Archiv, 1882, vol. xxxi. pp. 495, 501 (cases 4 and 10).

⁴ Neurol. Centralblatt, 1882, No. 17.

remarks made by Kast (loc. cit.). We have not to do with a fundamental departure from the typical course of R.D., but, apparently, with a mere deviation in those histological changes of the muscles which are doubtlessly at the root of the altered reactions. In the partial R.D. these alterations are usually insufficient to destroy the normal rapid wave of the neuro-muscular contraction; in certain cases they go so far as to change this contraction into a long-drawn one. The direct excitation of muscle by faradism is, we must remember also, of the same order as the indirect muscular one. This theory has nothing to militate against it, though of course it is not an easy task to give a histological proof of its truth. As Kast happily observes, our variety of R.D. may form a connecting link between the complete and partial forms.¹

I do not now think it so justifiable to call it "faradic R.D.," as Remak has proposed to do, since we find that indirect galvanic excitation may also be followed by long-drawn contractions. The main point, I repeat, is the occurrence of the abnormally slow muscular contractions to both kinds of electrical nerve-excitation, and to faradic muscle-excitation; hence I would propose to call this variety "the partial R.D. with indirect slow contraction" ("mit indirecter" oder "mit obligater," Zuckungsträgheit).

With reference to the clinical and prognostic meaning of this form, Kast is right in concluding from its occurrence to the presence of a change of moderate intensity and hopeful issue in the neuro-muscular apparatus. The cases in which it occurs may be expected then to run a favourable course.

These observations teach us that we have not yet exhausted the list of the numerous manifestations of R.D., and that the histological changes which are at the root of it may present numerous modifications of which we are as yet unable to give a connected account or explanation.

[A. DE W.]

¹ Cf. Erb, 'Elektrotherapie,' pp. 178 ff.; 197 ff.