sounds are scarcely complained of, and press these things into the wound. The drugs are used twice daily; the sounds with a frequency dependent on tolerance and individual need till repair is complete. Thus the wound is healed, mucous membrane at one end sending its epithelium to meet that from the other end, and over the sides as well. The desirable medicaments block dead spaces, and guard from infection.

(2) Guarding from urine contact is largely fulfilled by these, but I usually empty the bladder as often as necessary for the first two days. The best catheter I find to be a small rubber one stiffened somewhat by a slender whalebone stylet, having a coudée bend.

(3) As to complications, space forbids. Haemorrhage from before the triangular ligament is met by perineal pressure. A soda-water bottle filled with iced water fulfils a double function: it restrains priapism (often very injurious) and inflammation as well. It contracts the compressor bulbi urethrae muscle. Haemorrhage from behind demands pressure on the membranous urethra and a contracted bladder.

The difficulty of passing guides is met by the small device shown in Fig. 2. We can push but we cannot direct

FIG 2

or sustain guides. If introduced into the little cannula, they can be both directed and sustained. One secret is to fill the urethra with oil, then rotate and push.

I have operated very many times on the foregoing principles with results which justify me in bringing them to notice. I regret the illustrations are not quite so true as I would wish.

CONGENITAL HYPOTONIA (CONGENITAL AMYOPLASIA).

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THE main object of this paper is to describe a form of myopathy, exemplified by a patient under my care, which has apparently escaped observation in this country, at all events, although a condition similar in nearly every respect has been noted in Germany and America, and named "congenital hypotonia" or "myatonia congenita" by Oppenheim, the earliest and fullest writer on the subject. The patient was shown and her case discussed at a meeting of the Bristol Division of the British Medical Association, and many of the apposite suggestions and comments made there have been embodied in this paper.

E. G., a girl of 10, was brought to the out-patient department of the Children's Hospital, Bristol, on account of muscular weakness and general wasting. Her parents are healthy, and in a position to give their child good food and hygienic surroundings. They have lost three children since the birth of this one—one from measles, one from cancer of the mouth, and a third at two months old from some unknown cause. Her birth was uneventful and without instruments, at full term, and no relative, near or distant, has ever suffered from anything like this girl's malady. Within half an hour of her birth one hand was noticed to be peculiarly flabby and loose, but no other sign of weakness was apparent until she began to get about and to learn to walk. Then it was found that her legs gave way under her, letting her down upon the inner aspects of her legs in very much the same position as that which will be described later on; this prevented her from walking by herself till she was over 2 years of age. Her mother often found it very difficult to put the child's shoes and socks on, because the legs were so weak and flabby. When she was only 2½ years, the fact that her calves were very small was remarked upon; this feature has become more pronounced as she became taller and more definite of outline. Till she was 7 or 8, her ankles turned over so readily as to cause a serious disability, as she often fell down, and her attendance at school suffered many interruptions as a consequence. This trouble, however, has been greatly lessened by the use of boots with very long "uppers." Her hands and their possibilities have always been a source of entertainment to her friends; at school this has developed to such an embarrassing degree that her parents have been

obliged to forbid her "showing-off" to her schoolfellows. In spite of the muscular laxity, no dislocations have ever occurred. There has never been any defect of vision, and no squint has been noticed. Her voice does not disappear after prolonged use, and there has been no difficulty about mastication; it has, however, been noticed that she cannot cough powerfully. Her appetite is good, and she has never had any grave illness or serious injury. Her mental powers are quite normal. Her mother brought her to the hospital on account of the weakness of her hands, which was so great that she could not dress herself. She can hold up light objects, such as a cup of tea, but can do nothing with heavier weights—for instance, a bucket of water. Two miles is as far as she can walk at a stretch without being immoderately tired.

In appearance she is of average height; her face is not remarkably thin, and though the orbicularis palpebrarum seem slack and toneless, it is not possible to demonstrate weakness in any of the muscles supplied by cranial nerves. The muscles of mastication, together with those of the tongue and lips, are not appreciably atonic. It is when the upper limbs are examined that the most striking departures from the normal are found. There is a universal defectiveness of power, accompanied by, and apparently proportionate to, a poor development of the muscles—that is to say, the muscles of the hand and those controlling movement at the wrist are smaller and at the same time weaker than those of the upper arm. Unlike the muscular changes of acute and chronic anterior poliomyelitis and of most of the muscular dystrophies, no particular muscles or groups of muscles are selected by the process; the weakness and ill development are evenly distributed except for the fact already mentioned, that the distal parts of the limb have suffered more than the proximal.

Hence there is an entire absence of the contractures usually associated with muscular atrophy. This is also to be accounted for by the fact that, though the muscles are weak and ill-nourished, they are only relatively so; the grip, though very feeble, does exist, and similarly the muscles, though small and hypoplastic, are not completely wanting. Picked up in the fingers, they can be felt quite distinctly, though they are small, soft, and doughy; the impression given is different from that derived from finger atrophied muscles. These remarks, while applying specially to the musculature of the upper limbs, are true also of the general state of the muscles of the lower limbs and trunk, particularly of the former. The most striking feature of this enfeeblement is, however, neither the loss of bulk nor the loss of power, but the remarkable hypotonicity of the muscles; resulting from this is an abnormal freedom of passive movement about the various joints, which is well illustrated by the accompanying photographs. It is at the finger-joints and the wrists that the greatest mobility is found; each finger can be turned backwards till the tip touches the back of the hand just above the metacarpophalangeal knuckle, and similarly the hand can be extended on the forearm till the knuckles touch the back of the wrist. Flexion at the wrist is equally free; the palm of the hand can be pressed against the front of the wrist. The fingers can be spread out from each other widely; the hand can be rolled up into a cylinder with palm inwards and dorsum outwards, the tip of the little finger projecting beyond the ulnar border. The thumb, also, can be distorted into all sorts of fantastic shapes. At the elbow, recurvation is of course prevented by the olecranon process; but lateral movement is abnormally free, so that the forearm can be abducted till it forms quite a considerable angle with the line of the upper arm. The forearm can also be hyperpronated. At the shoulder there is nothing worthy of note.

The lower limbs are hypotonic, but not to the same degree as the arms; the gastrocnemii, like the extensors and flexors of the forearm, are small and soft. The toes can be doubled back at the dorsum of the foot; they can be drawn apart, and the foot can be crumpled up in much the same way as the hand. Movements at the ankle, also, are abnormally free; the peronei, for example, are lax, and readily allow of the displacement of the foot into an exaggerated equino-varus position. The knee affords a good instance of the fact that in this patient the mobility of a joint is limited, not by muscular tonus, but by the actual anatomy of the joint itself; recurvation is possible, but not beyond the point permitted by the ligaments of the knee-joint, wherein it seems to differ in degree from the recurvation sometimes seen in dorsal tabes, and called by Fraenkel "hypotonie musculaire." The patella can be laterally displaced to an abnormal degree. At the hip a very high degree of mobility is possible, abduction and adduction being allowed through a large sector of a circle; the freedom of internal rotability is attested by the remarkable squatting position assumed naturally and habitually by the patient. Instead of sitting on the floor with her feet in front of her, she rests her body, not only on the glutei but also on the postero-internal surfaces of the thighs, and on the inner surfaces of the knees, legs and feet, the knees being flexed; that is to say, the lower limb can be rotated inwards at the hip through an angle of 90°. Such a posture would be impossible if the outward rotators of the hip were possessed of a normal degree of tonus.

The muscles of the trunk have erred less widely from the path of natural development; yet there are very distinct indications of hypotonicity here also. The most notable of these

perhaps, is the exaggeration of the antero-posterior curves of the spine in the erect posture; in the lumbar region the spinal column bends forward so as to have quite a deep hollow above the sacro-iliac joints. This lordosis is similar in nature to that seen in cases of pseudo-hypertrophic muscular atrophy, and is, as in this latter disease, due to atony of the glutei, the erectors of the spine, and the muscles of the anterior abdominal wall. These latter are soft, even when the patient is doing her utmost to harden the abdominal wall; and, further, they bear a good deal more stretching than is possible in a normal subject, for she can bend backwards till her dorsal spine is at right angles to her thighs. That the erectors of the spinal column are feeble is proved by the forward stoop of the trunk when she sits on a chair. The spinati are small on both sides, leaving the scapular spines unduly prominent. The scapulae are winged, but not to the extreme degree often met with in the several forms of acquired dystrophy. The respiratory muscles seem to act naturally, and there is no disturbance of the functions of the sphincters.

Co-ordination is as good as might be expected, the gait showing no definite departure from the normal. I have observed no fibrillary tremor, neither is there any sort of coarser tremor to be seen. The knee-jerk is present on both sides, and brisk; there is a tendo Achillis jerk, but no ankle-clonus, and the plantar response is normal on both sides. I have not been able to elicit any of the usual tendon reflexes in the upper limbs.

The electrical reactions, which Dr. George Parker kindly investigated for me in the Electrical Department of the Bristol General Hospital, showed a definite decrease of muscular excitability equally to both currents, roughly proportionate to the diminution in volume and power of the muscles. The stimulability and conductivity of the nerve trunks were unaltered.

There is no diminution nor other perversion of sensation, special or cutaneous; and, apart from the condition of the muscles, there is nothing of the nature of trophic change.

With regard to the patient's progress, there is no doubt that any change that is taking place is for the better; massage seems to have produced a definite increase in the muscular strength of the legs, though the hands do not respond to treatment. According to her mother's description, the muscles have not grown flabbier since the early age at which it was first discovered that anything was wrong. It seems likely, therefore, that she may grow a little stronger in the next few years, but that she will ever have anything like a normal measure of muscular power is not possible.

What is the exact nature of this disorder? It has been present since birth, yet it can be ascribed to no inherited cause; no other case has occurred in any branch of the family, in which lies an important contrast to many of the other muscular dystrophies. That the loss of power is primarily muscular and not secondary to any central or peripheral nervous disease is shown by

(a) the fact that the muscles are weak in proportion as they are small and short; (b) the absence of sensory or trophic changes, fibrillary tremor, and reaction of degeneration. The small size of the muscles is apparently due, not to atrophy but to a failure of development which has been in operation through the child's life, and probably through the greater part, if not the whole, of the intra-uterine period of development; at any rate, it had begun

within half an hour of birth, and is at a standstill now. (c) A further point of difference from the type of muscular weakness associated with and dependent on changes in the nervous system is the diffuse distribution of the muscular affection; unlike focal and systemic diseases of the brain, cord, and nerves, the muscles affected are not separable into groups, but all the muscles of the body—except, perhaps, those of the face and eyes—are

implicated in the morbid process. This also is a point of difference from most of the other myopathies.

If, then, the nervous system is not at fault, is the disease entirely a muscular one? As was pointed out at the meeting where my patient's case was discussed, joints of normal configuration could hardly permit of such remarkably free passive movement; even in the presence of absolutely toneless muscles the wrist, for instance, could hardly be hyper-extended and hyper-flexed to the remarkable degree which is actually possible. The skiagraph which is reproduced seems to indicate a certain rounding-off of the articular surfaces, a deviation from the normal which would undoubtedly allow of passive movement through an excessively large arc of a circle. Not only the

joint surfaces, but also the other structures surrounding the joints—nerves, blood vessels, connective tissue and skin—must differ slightly from the normal in their anatomy, or pain and ischaemia would be produced by such a position as is for instance illustrated in Fig. 2. All these changes, or rather modifications of development, are, however, not primary; they would not have occurred if the muscles had been endowed with enough tone to prevent abnormally free passive movement. As it is, the muscular flabbiness has left the limbs more or less flail-like, and to this circumstance all the other structures have been compelled to adapt themselves.

The fault, then, lies primarily and almost entirely in the muscles themselves; the nervous system cannot be accused of complicity for the reasons detailed above. The case must be set alongside of the various forms of muscular dystrophy; from these, also, it presents certain definite diversities. To begin with, it is congenital and yet not familial in origin; moreover, the affection is spread diffusely over the whole body, and has not selected any particular muscles or groups of muscles; there has never been any pseudo-hypertrophic stage, and the process is at a standstill. Having, therefore, ruled it out of court not only the ordinary myopathies, but also central nervous disease, we yet have to see whether the case comes under the head of any disease hitherto described. At first I

was inclined to suppose that it belonged to no recognized group, but further examination of medical literature shows that in nearly all details the case corresponds to those described first by Oppenheim under the name of "congenital hypotonia" or "myatonia (not myotonia) congenita" in two separate papers,¹ as well as in his textbook of nervous diseases.² The only other writer on the subject is Spiller, whose record of a case includes the only description

Fig. 1.

Fig. 2.

of an autopsy hitherto performed upon a subject of this malady. Briefly, Oppenheim brings forward as a definite clinical entity a general flaccidity of the skeletal muscles, excepting those supplied by cranial nerves, noticed at or soon after birth. It varies in degree; in extreme cases the limbs are mere useless appendages. The distribution of the muscular affection also varies. In one case all the limbs are affected, in another the legs alone, in another the muscles of the head and trunk suffer, but not those of the limbs. The muscles are not atrophic in the usual sense of the word, but soft and atonic; they react feebly or not at all to mechanical and electrical stimuli. Afferent paths are unaffected, the mind appears normal, and the sphincters act well. Oppenheim is inclined to give a hopeful prognosis based upon the course of the four or five cases he has seen. As he points out, the congenital nature of the complaint, the absence of the familial factor, the generalized distribution of the muscular change, and the tendency of the patients to get better rather than worse, separate it from acute and chronic anterior poliomyelitis, multiple neuritis, the various myopathies, and the pseudo-paralysis accompanying syphilitic, scorbutic and other diseases of the epiphyses. In his opinion, the fact that no history of injury at birth was forthcoming in any of his cases suffices to annihilate any theory of haemorrhage into the medulla or cord. He had met

with no cases except in infancy. The only other description is Spiller's,³ whose patient was also an infant; death was apparently due to tuberculosis. Clinically the case coincided exactly with Oppenheim's account. *Post mortem*, the affected muscles were found to be small, pale, and "hyaloid"; the subcutaneous fat constituted an unduly large proportion of the bulk of the limbs. The intramuscular connective tissue was excessive in bulk and in the amount of fat it contained; the muscle fibres were small, in the limbs especially,

and to a less extent in the trunk, and had undergone a patchy hyaline degeneration. There was absolutely no demonstrable change, macroscopic or microscopic, in brain, cord, or nerves. Certain changes were noted in the thymus⁴—a point of interest when conjoined with the fact that a persistence of the thymus is sometimes associated with myasthenia gravis. There was general fibrosis of the gland, with increase in the number and size of Hassall's corpuscles; in both thymus and spleen there was evidence of proliferative change in the intima and adventitia of the arterioles. Other points noticed were the presence of lymph nodes within the lungs, and proliferation of the lymphoid tissue of the mesenteric glands. Whether these changes bear any real relation to the nature of the disease is very doubtful, since the functions of the thymus are so little understood; fibrosis of the organ has been shown by my colleague, Dr. J. M. Fortescue-Brickdale,⁵ and others,⁶ to be commonly associated with idiopathic infantile atrophy.

My patient shows certain differences from the above descriptions; neither of the writers mentioned has seen a case of more than 2 years of age, but probably some of Oppenheim's patients, if they live to be as old as mine, will show a precisely similar condition of the musculature. The limbs in his cases were not small, while in my case they are very thin; this difference is to be

accounted for by the relative excess of subcutaneous fat in infancy over the amount present in later years. In Oppenheim's cases, and in Spiller's, the tendon reflexes were absent; but, in view of the difficulty of obtaining reliable evidence with regard to these phenomena in a child, this discrepancy is of little importance. In all other points the similarity is complete, and there can be no doubt that my patient exhibits the same condition, persisting till the age of 10, which has so far only been recorded as existing in babyhood. Since this patient was shown at the meeting alluded to above several people have recollected similar accomplishments among their friends and relatives, but on a less ambitious scale; and it is possible that many of the "double-jointed" children who are always in great request among their school fellows, and the "boneless boys" of travelling circuses, are "formes frustes" of this curious muscular disease.

The term "amyoplasia" has been suggested as a substitute for "hypotonia" because the latter only expresses that feature of the disease which most struck Oppenheim; it is desirable to find a name which embodies that fuller information with regard to the nature of the malady which has been furnished by Spiller's autopsy, and the case recorded here in a patient of an age at which it is easier to appreciate the nature of a muscular disease than it is during infancy.

In conclusion, I must acknowledge my indebtedness and express my thanks to Dr. Newman Neild, Assistant Physician to the Bristol General Hospital, and to Mr. Ellington Jones, Skiagraphist to the same institution, for the excellent photographs and radiographs which they were good enough to take for me.

POSTSCRIPT. — To the foregoing summary of this interesting disease must be added a brief allusion to several cases published recently by Italian physicians. Unfortunately the descriptions available are meagre. Sorgente⁷ contributes an account of two

cases, both of them infants under a month old, with a general flaccidity like that which Oppenheim described; the thoracic muscles were affected as well as the limbs, which probably accounts for the death of both from bronchitis. In both the electrical reactions were doubtful and the knee-jerks brisk. Sorgente alludes to two other contributions to the subject without giving precise references; Muggia of Turin described a case in 1903, and Berti of Bologna in 1904 published an account of two more. In none of these was there any autopsy. Another paper by Jovane of Naples⁸ describes a case in a child of 3 months, who died of bronchopneumonia, a calamity favoured by the fact that the thoracic muscles did not escape the flaccidity which was distributed throughout the limbs and trunk. In all essential points the case was like the others summarized here; the tendon-reflexes were absent, unlike those in Sorgente's cases and my own. There was no *post mortem* examination, nor any fact in the history that could throw light on the etiology. From these Italian cases the immediate prognosis would appear to depend on the state of the thoracic muscles; if these are involved, the infant is likely to die of bronchitis.

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Fig. 3.