ARACHNODACTYLY.

BY

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The first description of arachnodactyly dates back to 1896.

In that year Marfan⁹ showed a case of a child exhibiting unusual length and slenderness of the bones of feet and hands, and pointed out that other abnormalities coexisted; he suggested the term 'Pieds d'araigne' (Spider feet) but decided upon that of 'Dolicho-stenomélie'; the possibility of endocrine disorder was first recognized by him, but he was unable to substantiate his theory. Six years later Marfan's case was shown by Méry and Babonneix¹⁰ who showed by means of skiagrams that some activity of epiphyseal cartilages was present, and used the term 'Hyperchondroplasia'; they also mentioned syphilis as a possible factor. In the same year (1902) Achard¹, using the term 'arachnodactyly' for the first time, demonstrated the condition in a girl aged 18 years, where both familial and hereditary influences were marked. In the following year Poynton¹⁵ showed a case to the Medical Society of London under the heading of 'Atavism,' which seems without doubt to have been one of arachnodactyly. On this occasion a member present at the meeting suggested that the condition might belong to the group of muscular dystrophies. In 1914 Borger² described two cases in Germany, and laid stress upon its likeness to acromegaly. Salle²¹ in the same year spoke of a combination of giant growth and arachnodactyly. Thursfield²⁴ in 1917 favoured the dystrophic view. He drew attention to the atony and ill-development of the entire musculature of body and limbs, and laid stress upon its resemblance to amyotonia congenita. In the same year Rietschel¹⁸ described a case in which the orbital fissures were obliquely placed, and therefore likened the condition to mongolism. Poynton and Maurice¹⁶ in 1913 demonstrated an instance in which the onset was believed to start in later life, at the age of 6 years; they favoured the endocrine rather than the dystrophic theory. In 1924 Ormond and Williams¹² quoted the details of a case with special reference to ocular symptoms. The first case to be described in America was that by Piper and Irvin-Jones¹³ who regarded congenital heart disease as a prominent feature of arachnodactyly, and described the post-mortem appearances of a case in which this was present. Schlack²² in 1926 recorded the only instance with symptoms which suggested definite pathological changes in the nervous system.

From a review of the literature it seems evident that the condition is a definite clinical entity, and although various theories of the causation have been suggested no one of them is universally accepted.

Only twenty-two cases appear to be on record. Four cases are described in this paper. One is an undoubted instance of amyotonia congenita; in another the atonicity of the musculature is less striking but evident. The remaining two cases are, I think, unique in that they are brother and sister. Their mother was a similar instance of arachnodactyly, and moreover all three members of this family showed many other striking congenital abnormalities.

ÆTIOLOGY.

Various theories of the causation of this condition have been mentioned above. These may be conveniently examined under their separate headings.
(a) *An endocrine disorder.*

Consideration of recorded cases and those of my series shows that with perhaps two exceptions the onset of arachnodactyly takes place during intra-

uterine life. Of 26 cases, in 16 the condition was noticed at birth. In one (Poynton) it was believed to have started later, and in one of my series (Case 1) it was not noted until some years after birth. In seven cases there is no mention of the date of onset, but of these, six showed obvious congenital abnormalities and the remaining case suggests an onset before birth.

This occurrence in foetal life is of great importance, for we are not accustomed to see at birth abnormalities such as in the older child are recognized to be of endocrine origin: indeed, it is stated that any perverted function of these glands cannot affect the foetus, as such deficiencies are compensated for by the mother. It has been thought that perhaps arachnodactyly might be a foetal gigantism of pituitary origin, the result of over-secretion of this gland. If we consider the three cases in which hereditary influences were marked we might argue that increased maternal secretion had been handed on to the child *in utero*, but in all these three instances other members of the family escaped: moreover, the mothers of these children did not show signs or symptoms known to be the result of such increase—they were not instances of acromegaly. Partial gigantism in the child persisting as in these cases into adult life without the development of true acromegaly would suggest that the activity of the morbid process ceased before, or at any rate shortly after, ossification was complete, and we should need to invoke such an explanation if we regard these cases of pituitary origin, for in the records of those examined in later life no signs or symptoms of acromegaly were found to be superimposed. We might suppose, perhaps, that during intra-uterine life the foetus receives an abnormally increased amount of pituitary secretion, but that after separation from the mother it then depends upon the normal amount supplied by its own gland; and that thus the morbid process, removed at birth, cannot later produce acromegaly.

The enlargement of the lower jaw, the prominence of supraorbital ridges, the hypertrophied ears and many other signs, including even the muscular weakness, lend some support to the theory of pituitary hypersecretion; but it must be remembered that arachnodactyly is a partial gigantism only, for although the length of the complete skeleton is frequently increased, the elongation of the phalanges of hands and feet is relatively still greater. Moreover, it is most difficult to account for the thinning of these bones which has been found both by X-ray and post-mortem examination: this is directly opposed to the findings in gigantism or acromegaly in which the breadth is actually increased. Evidence afforded by autopsy is conflicting. Salle²¹ found enlargement of the sella turcica, with the presence of a bony tumour growing out from the base of the skull in such a way as to press upon the hypophysis, which was, however, normal on microscopical examination. Borger², on the other hand, stated that in his case he found enlargement of the sella turcica posteriorly, thickening of the clinoid processes and flattening of the hypophysis: microscopically the posterior lobe was normal but the pars anterior and pars media were cystic, the epithelium was cubical in type, and large numbers of eosinophil cells were present.

My own clinical observations do not suggest that the pituitary is at fault. The gigantism is partial only, as in recorded cases, and though there is no marked thinning of the bones the breadth is certainly nowhere increased. Other features connected with pituitary disturbance are absent: none of the cases complained of headache, nor was optic atrophy present in any of them; no degree of temporal hemianopia was found, and the colour vision was normal in each case. Nor are there any signs or symptoms which might point to a hypo-secretion, the low systolic blood pressures and the decreased basal metabolic rate in one case are to be expected in such weakly children. On the other hand, by X-ray examination of the skull certain interesting features were noted. In each radiogram some change in the sella turcica was seen. In Case 3 it was not only small but appeared elongated, both anterior and posterior clinoid processes were thickened, but the opening was very little reduced in size, owing possibly to the general shape of the skull. In Case 4 the sella turcica was again smaller than normal and with a similar thickening of the clinoid processes, the opening was small but there was no elongation. In Case 1 a very marked reduction in size of the sella turcica and clinoid processes was seen. The radiograms did not appear to show any other abnormalities.

These appearances suggest that the pituitary body is smaller than normal, but in the light of clinical findings it does not seem that its function is in any way perverted. Lastly, the coexistence of so many varied congenital stigmata is unfavourable to the theory.

The same points apply with regard to other endocrine glands; these cases do not suggest an abnormality of thyroid or other secretions.

(b) *A condition allied to mongolism.*

This suggestion seems to have a very doubtful foundation. Certainly abnormalities such as the highly arched palate, laxity of joints and ligaments, and especially deformities of the heart, are common to both, but arachnodactyly differs strongly from mongolism in other respects. The mentality is excellent. The head is commonly dolichocephalic whereas in mongolism it is brachycephalic and usually devoid of eminences. The mongolian hand differs markedly with its thick fingers rounded at the tips, short thumb and dwarfed little finger, and the characteristic large fissured tongue of later years is not seen in arachnodactyly. The oblique orbital fissures of the mongol have certainly been noticed in one instance by Rietschel, but the changes in the extremities of the bones found in arachnodactyly are unknown in mongolism. With regard to family history, the mongol is usually looked upon as an exhaustion product, a large proportion of the cases coming at the end of large families and being the offspring of mothers near the end of their child-bearing period; this does not seem to be so in arachnodactyly, with one notable exception amongst my series, (Case 1), who was the last child of a family of fifteen and whose mother was forty-two years of age at the time of the patient's birth.

Fig. 1. Case 1. Showing the large lower jaw. The features here are very old for the age.

Fig. 2. Case 1. Showing the long slender and somewhat tapered fingers. The appearance of muscle wasting may be seen.

Fig. 3. Case 3. Showing the clubbed feet and deformed toes.

(c) *A form of hyper-chondroplasia.*

This term was originally suggested by Méry and Babonneix because skiagrams showed some activity of epiphyseal cartilages, but in later cases this has not been a feature, and in my series the epiphyses were normal. Nor can there be any connection with rickets though, as in the case quoted by Thomas²³, the two diseases may coexist; the early general indications of the latter are not seen in arachnodactyly, in which, moreover, dentition is normal.

FIG. 4. Case 1. Showing the increased length of the phalanges; tapering is slight. No thinning nor rarefaction of bone is seen.

(d) *A form of primary muscular dystrophy.*

This suggestion was first advanced by a member present at the Medical Society of London in 1903 and again in 1917 by Thursfield who was the first to lay stress upon the atony and ill-development of the entire musculature.

That arachnodactyly is closely allied to amyotonia congenita is, I think, certain, though there appears to be no connection with other dystrophies. The comparison here made is chiefly based on Reuben's¹⁷ account of 6 cases of amyotonia and a review of 136 others. Hereditary and familial influences are interesting. In 3 out of 26 cases of arachnodactyly both factors are

FIG. 5. Case 1. Showing the curious dwarfing of the middle phalanges of the little toes.

strongly marked, no mention is made in 6, and in the remaining 17 the histories are normal. Amyotonia congenita is also occasionally hereditary but more commonly familial, though Kerley and Blanchard⁷ state that neither influence is usual. With regard to sex incidence, in arachnodactyly there were 10 males and 16 females, *i.e.*, a slight preponderance (61%) of the latter, whereas in amyotonia there is a corresponding increase in the male sex. The onset in

both is almost invariably intra-uterine, though amyotonia has been known to occur at a later period. The mentality found in the two conditions corresponds, being as a rule excellent. Congenital abnormalities and those resulting from muscular weakness occur in both, though in amyotonia they are comparatively rare: of these may be mentioned deformities and asymmetry of the skull, winging of the scapulæ, dislocations of the hip, club foot and various deformities of the thorax. Kyphosis and scoliosis are commonly found in both. One of

FIG. 6. Case 1. Showing the small sella turcica and clinoid processes; the latter are somewhat thickened.

the most interesting congenital abnormalities which occurs in a high percentage (27%) of cases of arachnodactyly, namely, congenital dislocation of the lens of the eye, is however not recorded in amyotonia by Reuben or other writers.

The great feature which these two conditions do undoubtedly share is the flaccidity of the musculature and to a slightly less extent relaxation of the ligaments. Of the 26 cases arachnodactyly there is mention of the muscles in no less than 19, which in 17 showed apparent atrophy. Laxity of the ligaments is a feature in 18 instances and not mentioned in 8. Resulting contractures

were present in 14. These figures may easily be too low, for in some of the recorded cases it is likely that no heed was given to the muscular system. It may also be noted that in the 9 cases in which some degree of muscular atrophy is not recorded, only 4 observers fail to mention the muscular system at all. Of the remaining 5, one (Fowler) had contractures of the forearm and marked hypotonicity, another (Rocher¹⁹) had contractures of the hamstrings and loose articulations. Achard¹ states that in his case the muscles were normal and contractures absent, but he does note an exaggerated mobility of joints, Pfaunder¹⁴ speaks of a slenderness of soft parts and Lust's⁸ case had flexor contractures of the fingers and toes. The muscles are therefore indirectly alluded to in a further 3 cases, and in yet another a 'slenderness of soft parts' seems suggestive. Thus the muscular system would appear to be affected in no less than 80% of total cases.

With regard to electrical reactions, Marfan⁹ found a normal result but Dupérie, Dubourg and Guénard⁵ noted a diminished response to faradism. Of my cases, Case I, showed a slight but equal diminution of response to both faradic and galvanic currents, though a contraction was always obtained even from those muscles which appeared most wasted. In the other two cases the reactions were in every respect normal.

Organic disease of the nervous system is absent in both arachnodactyly and amyotonia, though Schlack in his account of a case of arachnodactyly described signs suggestive of affection of the posterior columns and pyramidal tracts, likening the case to Friedreich's disease. It is difficult to know in this sole instance whether such nervous phenomena are in any way related to the congenital abnormalities.

On the muscular findings alone arachnodactyly must surely be thought to possess one causal element in common with that of amyotonia, though the presence of some other factor or perhaps the selective action of this may be of importance. It is not suggested here that the two conditions are one and the same, but that there exists an exceedingly close relationship in the matter of ætiology; it is believed that an amyotonic state of the muscles is a constant accompaniment of arachnodactyly though the degree in which this feature is present varies. A combination of the two conditions each strongly represented in one patient has not been hitherto recorded, but two such instances are described later in this paper.

(e) *A fault in the embryo.*

Borger², in discussing the ætiology of arachnodactyly, has spoken of 'a partial gigantism caused by defective anlage or faulty predisposition of the entire organism, or perhaps an early nutritional defect of the embryo.' It has been shown that the condition starts during intra-uterine life, and it is interesting to note that dislocation of the lens is such a frequent feature of arachnodactyly, for this abnormality is believed to occur within the first three or four months of foetal life. To form an opinion on this question is difficult. If a faulty predisposition is handed on to the embryo the condition becomes a matter of heredity and would probably be familial as well, but in

three cases in which these influences were present there were children in whom the condition did not appear: in each instance the heredity was entirely on the female side. Dupérie, Dubourg and Guénard⁵ state that in their case the parents were very young, the father being only seventeen and the mother sixteen. In one instance (Marfan) a shock of an hallucinatory nature occurred early in the pregnancy. One case was four weeks premature and an only child. Miscarriages are noted by Marfan⁹ and Salle²¹, but for the most part confinements were normal. With the exception of one of my cases (Case 1) the pregnancies do not seem to have occurred either late in life or to have succeeded many others, which is opposed to the theory of a reproductive exhaustion put forward by Nobel¹¹; this girl however, was the last child of a family of fifteen, her birth followed upon six consecutive still-births and the mother was forty-two years of age at the time. Tests made to determine any syphilitic infection were invariably negative.

Thus, beyond the hereditary factor and the occurrence of the condition in more than one but not all members of the families, there is little to note. Damage of an environmental kind must be supposed to be the result of some nutritional defect, physical or chemical in nature, such as could be brought to the embryonic tissue in the maternal blood, but in the large majority of cases the condition does not occur in more than one pregnancy and therefore the defect must be a temporary one. Recently much interest has been taken in the condition hypertelorism, first described by Greig, of which the main feature is an abnormal distance between the eyes. Though this peculiarity is not found in arachnodactyly there are several features common to the two conditions, such as the preponderance in the female sex, the congenital factor, arching of the palate, prognathous jaw, small mouth, large ears and other deformities. It is particularly interesting to note that in one case described by Braithwaite³ the fingers and toes were thin and tapering and muscle tone was markedly deficient, recalling the contortions of amyotonia congenita. In hypertelorism also hereditary influence is occasionally though rarely evident. Although the two conditions differ in certain clinical manifestations, the fundamental factors in their ætiology must be closely allied.

Further observations upon ætiology.

Certain investigations were made in my series of cases but mostly yielded negative information. Renal function was found to be healthy; the urine by chemical, microscopical and bacteriological tests was normal, the blood urea was satisfactory and the concentration of this substance was excellent. The urinary diastase test gave normal results in two cases, but in the third (Case 1) the content was found to be 90 units. Although this figure proves that the ability of the kidney to excrete this ferment is good, it seems to suggest a hyper-secretion on the part of the pancreas, assuming that this organ is responsible for its secretion, a view which has been criticized. From a complete examination of the stools the pancreatic functions appeared to be normal and it is difficult to account for this high diastatic index. The liver also was healthy. Total and differential blood counts were normal. A Sachs-Georgi

reaction carried out in Case 1 showed no evidence of syphilis. The calcium content of the blood was normal. The decreased basal metabolic rate in this case has been mentioned. Blood pressures were found to be low; pulse pressures were high. The electrical reactions of the muscles have been described.

Of the various investigations made the only unusual findings obtained were those in connection with the sella turcica, and these have been discussed in that portion of the ætiology which deals with endocrine glands.

FIG. 7. Case 3. The length of the phalanges is increased. No further changes are apparent.

CLINICAL FEATURES.

The onset during intra-uterine life and the preponderance in the female sex have already been mentioned. It is convenient first to give from a survey of the literature the main clinical features to which the term arachnodactyly has been applied, then the observations on those points from my series of

cases, and lastly to discuss the many accompanying abnormalities under their respective headings.

A review of the literature shows that the striking characteristic consists of an abnormal lengthening of hands and feet especially noticeable in the fingers and toes. This is demonstrable not only as a real increase by comparison with children of the same age, but also as a relative increase by comparison with the total length of the child, although this is usually itself increased.

FIG. 8. Case 3. Bones are increased in length. No thinning nor rarefaction is seen.

Thus the child is abnormally tall for its age with hands and feet relatively long for its height. Equally striking is the thinning of the bones involved, and in the fingers especially there is frequently a tapering towards the distal extremities of the terminal phalanges resulting in a spidery appearance to which the term *arachnodactyly* is appropriate. The length and slenderness of the fingers are accentuated by a deficiency of the surrounding soft parts. The metacarpal and metatarsal bones share these peculiarities in many but not

all cases, and if so are usually affected to a lesser extent. The arms may escape, and any increase in length in them tends to be relatively greater in the forearms than upper arms, the elongation becoming more marked towards the distal end of the limb. The same is true of the legs.

Fingers and toes are not clubbed, though Thomas²³ notes this feature. Joints are very freely movable but structurally normal, the limbs are hyper-extensible without pain. Webbing of the fingers is not uncommon. Two cases of talipes calcaneus are described and one of hammer toes (Dubois⁴). Pes planus is common, pes cavus less so. Spurring of the os calcis is noted in 13 or 50 per cent. of total cases.

Whereas the height tends to be greater than the average for the age, the weight is considerably less. This is due to the deficient musculature and to the loss of subcutaneous fat. The consistency of the muscles is much altered and suggests that of adipose tissue.

My own observations differ in some respects from the foregoing description. In two of my cases there was a marked increase in height with a slight increase in weight also, and in two an equally marked decrease in both height and weight. Measurements were taken in each case of the lengths of the spine, upper arm, forearm, thigh and lower leg, and compared with those obtained from the average of a series of normal healthy children of the same age and sex (see Appendix, Table A). In the cases in which total body length was increased the figures obtained were in excess of the normal for the age, but where body length was decreased, they were smaller than the normal. The ratio of each measurement to the corresponding total height was therefore estimated. The results were very consistent and showed that the ratios were undisturbed, corresponding accurately with the normal (Table A). From this it is clear that in the long bones of the skeleton there is no progressive increase in length, for where elongation occurs in these bones it is shared equally amongst them and is in normal proportion to the corresponding increase in the height. There is therefore no partial gigantism.

In order to investigate the condition of the bones of the hands measurements were taken in millimetres from skiagrams of every phalanx and metacarpal in each of the cases; these were then compared respectively with similar measurements obtained also from skiagrams of normal children of corresponding ages (see Appendix, Table B). It was found that there was a definite increase in length in every measurement taken with the exception of two distal phalanges and three metacarpals in the case of Case 1. It was also noticed that the figures obtained in Case 4 at the age of $7\frac{1}{2}$ years were mostly equal to, or greater than, those of the normal at the age of 14 years with which Case 1 is compared. This is so remarkable that these figures at 14 years are given in comparison with this case instead of those of a corresponding age. Individual discrepancies are obviously much greater in the two cases in which body length is increased, but even in those in which this is decreased there is a noticeable difference; the figures shown in Table B are, I think, very striking, especially the comparison of the sum total of the phalanges and metacarpals with the normal. Since the figures in the cases in which height is decreased still show

an increase compared with the normal, it is obvious that the elongation in these bones is out of proportion to the body length. In each case ratios of the total sum of the phalanges and metacarpals of the second or longest digit to the respective body lengths were estimated and compared with the normal (Case 4 is again compared with the age of 14 years). The results prove that these bones exhibit a true partial gigantism (see Appendix, Table C).

In order to determine whether the elongation was confined to the phalanges or shared by the metacarpals, I estimated the ratios of the sum of the phalanges of each digit to the sum of the phalanges plus corresponding metacarpal bones in every case, and compared these with the normal (see Appendix, Table D). In Cases 3 and 4 (in which body lengths were increased) it was found that the phalanges of every digit were relatively longer to their corresponding metacarpal bone than in the normal child of the same age, the exact ratios being almost identical in these two children. In Case 1, however, the sum of the phalanges appeared to show a slightly smaller proportion of the whole, *i.e.*, the metacarpals were relatively longer. In the remaining case (Case 2), some variation was observed, but on the whole the increase seemed to be shared equally. Finally, taking the second digit only in each child, the ratios between individual phalanges and their corresponding metacarpals were estimated (see Appendix, Table E). The figures show that the greatest relative increase lies invariably in the terminal phalanx. In Case 1, both middle and proximal phalanges were also increased relatively, but in the other cases there was some variation.

With regard to the feet, to obtain measurements accurate enough to draw conclusions is practically impossible. Owing to accompanying deformities, such as talipes, foreshortening appears in the photographs and the proximal ends of metatarsals especially are frequently blurred. Appearances suggest that a similar state of things obtains as in the hands.

By X-ray examination it was also observed that the thinning of the bones was more apparent than real, nor is tapering very evident, the deficiency of soft parts is probably largely responsible for the spidery appearance. No rarefaction was observed nor was there any evidence of advancement of ossification.

As in recorded cases there was a marked freedom of movement at the joints with a laxity of ligaments. The muscular condition was striking. In two cases the apparent wasting was general in distribution with the exception of the face which entirely escaped, but the loss of power was not in proportion though Case 2 was a very weakly specimen.

In addition to the peculiarities of hands and feet, a host of abnormalities in other parts of the body has been described, many of which I have observed in my series of cases.

Mentality. This is mentioned in 19 cases, of which 17 (90%) are normal. In two of my cases it was impaired during the first few years only.

Teeth. In two cases dentition was delayed, early caries was noted in three and in one (Rominger)²⁰ there was a double row of teeth in the lower jaw. The teeth are mostly normal.

Speech. Occurred late in 4 cases, some impairment in 4 others.

Skin. Mentioned in 11 cases, in 3 described as pale, in 3 as dry and fine, in 5 there was no peculiarity.

Hair. Described variously as being thick and dry, coarse, straight, and abundant. No type is peculiar to the condition.

Head and Face. Abnormalities of the skull are extremely common, being present in 17 or 65% of all cases. In 6 the shape was found to be of the dolichocephalic variety, in 1 only it was brachycephalic. Other deformities found

FIG. 9. Case 3. Showing a curious elongation of the sella turcica; the clinoid processes are thickened; the opening is very little reduced in size.

are :—Asymmetry in some form in 3, relative smallness of the skull in 2, prominence of the chin in 5, prominent supra-orbital ridges in 4, bossing of the frontal eminences in 5, patency of the anterior fontanelle in 2, depression of the base of the skull in 1, prominence of the nose in 1, poor development of the base of the nose in 1, and of the inferior maxilla in 1. There was no mention of the skull or head in 5 cases, in 4 it was described as normal. The face was mentioned in 8 cases as being old for the age, the lips small in 4, the whole face small in 1, in 1 the nose was large and in 1 the neck was short. Marked naso-labial folds were also noted, the oblique axes of the orbital fissures of mongolian type were present in 1 case. Salle mentioned a large tongue. The

vault has been found to be thin at autopsy, and in 2 cases the roofs of the orbits projected backwards unduly into the interior of the skull.

Thorax. Abnormalities are present in 13 or 50% of cases; the funnel shaped chest was found most common by occurring in 6 (23%), projection of the lower sternum with flattening of the whole or part of the chest wall in 3. pigeon breast was also seen. Rarefaction of the ribs was described in 1 case.

FIG. 10. Case 4. The increase in length of the phalanges is very marked and actually bears comparison with Case 1 who is $6\frac{1}{2}$ years older.

Scapula. Described as winged in 4 cases, long in 2, rarified in 1, normal in 4. Not mentioned in 16.

Spine. A kyphosis alone is common (45%), but there may be a scoliosis or a combination of the two. These are probably always the result of muscular weakness. In 1 case the pelvis was asymmetrical.

Palate. This was noted as highly arched in 10 cases. In 1 there was a median cleft in the hard portion. It is possible that the highly vaulted palate

occurs more often than the figures suggest since it is an abnormality which has specially to be looked for.

Ears. Deformities were mentioned in 17 cases all of which affect the external ear only. Most common were a general enlargement of lobes especially deficiency of cartilage, and over-development of the crus helicis. Less common are over-development of the anti-helix, tragus, and cartilage, depressions in the concha and bad definition of the crus helicis, anti-helix and lobules. A

FIG. 11. Case 4. The bones show some increase in length. No other abnormality is present.

division of the lobules into separate portions has also been described. A combination of abnormalities is usual.

Eyes. These are amongst the most interesting of all accompanying congenital deformities. Not only is their occurrence in some form or other very frequent, being noted in 50% of all cases, but in addition one particular abnormality, that of dislocation of the lens, occurs most often. In conjunction with other congenital maladies of the eye this condition is extremely rare, yet it is in arachnodactyly a feature in fully half of those cases which show ocular abnormalities. Of 26 cases the eyes showed some peculiarity in 13, and of

these 7 had luxation of the lens. In many the eyes have probably escaped examination, in 9 they are not mentioned, and once again it is possible that the incidence of this particular feature is higher than the figures suggest. It is also interesting to note that dislocation of the lens never occurred as a single abnormality. It was accompanied by iridodonesis in 4 cases, in 2 of which congenital hydrophthalmos was also present. The pupils were frequently very small and the globes deeply set. Squint occurred often, nystagmus

FIG. 12. Case 4. The sella turcica is reduced in size; clinoid processes are thickened.

occasionally. A persistent pupillary membrane was mentioned in 1 instance. High degrees of myopia occurred, and amblyopia, enlargement of the cornea, and shallowness of the anterior chamber were recorded. Optic discs apart from myopic appearances were normal; no instance of congenital cataract was found. Accompanying peculiarities such as œdema of the eyelids, deficiency of eyelashes, marked epicanthic folds, and variations in the size of the two orbital fissures were also mentioned. Rietschel in his description of the

slanting axes of mongolian type made no further reference to the eyes. A list of the various ocular abnormalities, showing their total and relative incidence, is shown in Table I.

TABLE I.
OCULAR ABNORMALITIES IN ARACHNODACTYL (26 CASES).

Abnormality	No. of cases	Total incidence	Relative incidence
Dislocation of the lens	7	27%	54%
Iridodonesis	5	19%	38%
Small pupils	5	19%	38%
Deeply set globes	4	15%	30%
High myopia	3	11%	23%
Squint	3	11%	23%
Nystagmus	2	8%	15%
Congenital hydrophthalmos	2	8%	15%
Enlargement of the cornea	1	4%	8%
Shallow anterior chamber	1	4%	8%
Persistent pupillary membrane	1	4%	8%
Amblyopia	1	4%	8%
Fundus oculi	Normal except for myopic appearances.		

Total number of cases, 26. Numbers showing abnormalities of the eyes, 13 or 50%. Number in which there is no mention, 9.

Lungs. At autopsy Borger found a deficiency of the middle lobe of the right lung, and in the left a large lingula was present. Piper and Irvin-Jones found a very small middle lobe in the right lung and the left consisted of one lobe only. Salle's case showed no defect. Death was due to pneumonia in each instance. The liability to this disease is great since the bony thorax is so frequently deformed.

Heart. Affections of the heart were mentioned in 9 cases, of which 5 are considered to be congenital in type. In 2 a patent foramen ovale was seen at autopsy and in a third the interauricular septum was deficient. Valves were normal in each case. Amongst my series no abnormalities were found.

Viscera. Salle found a splanchnomegaly or increased length of the gut. With this exception the viscera are normal. No defect of the abdominal wall was recorded.

Ductless glands. Apart from Borger's account of the pituitary body and the X-ray findings of the sella turcica in my cases, both of which have been mentioned, there is no evidence of abnormality of any ductless gland. Borger also described the external genital organs as small in one instance, but in another the mons veneris and labia were very large, a feature noticed by Pfaunder.

Nervous system. With the exception of Schlack's account which is described above (see *Ætiology*, section *d*) no abnormalities of this system are on record.

DIFFERENTIAL DIAGNOSIS.

This presents no difficulty if the hands and feet are examined, the symmetrical involvement and the absence of bony disease will exclude other conditions. It may be distinguished from infantile paralysis, obstetrical and diphtheritic palsies by the absence of organic changes in the nervous system. In cases of amyotonia congenita, in which the muscular condition may completely dominate the picture, an accompanying arachnodactyly might remain undiagnosed unless the possibility of its existence be remembered. Wasting may suggest tuberculosis which may, of course, be present at the same time. The points which should distinguish mongolism and acromegaly have been mentioned. Syphilis is excluded by the Wassermann or other test.

CONCLUSIONS.

1. Arachnodactyly is a clinical entity of rare occurrence. It is congenital in origin, and its onset probably takes place during the early months of intra-uterine life. It is more common in females than in males in the ratio of 3 to 2.
2. Heredity is an important factor in causation though apparently absent in the majority of cases. The condition is not usually familial though it may occur in more than one member of a family.
3. It is not a result of parental reproductive exhaustion though one case suggests this possibility; neither is there in this respect nor from clinical findings a connection with mongolism. Pathological evidence shows that it is not a form of hyperchondroplasia.
4. Radiograms showing abnormal appearances of the sella turcica are described in three cases, but since no clinical evidence suggests a perversion of pituitary secretion the view is held in this paper that the functions of the pituitary are normal, nor are there grounds for supposing an abnormality of any other endocrine glands.
5. The functions of those organs especially connected with metabolism such as the liver and pancreas, and with excretion such as the kidney, have been investigated. No defects of any sort were found.
6. Clinically the condition exhibits a true partial gigantism, as is proved by a comparison of measurements obtained by X-ray photographs with the normal. The increase in length is present especially in the bones of the hands, of which the terminal phalanges are the most affected.
7. The characteristic deformities of the hands and feet never occur alone, other congenital stigmata are always to be found. The frequent occurrence of abnormalities of the eyes is emphasized, particularly that of congenital dislocation of the lens.
8. An atonic state of the muscles is constant and may be regarded as a part of the condition. This conforms in all essentials to that found in amyotonia congenita though it varies greatly in degree in different patients. It is suggested here that though the two diseases are not one and the same, the fundamental factors in the causation of both are very closely allied.

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(For Appendix, see p. 210).

APPENDIX.

AUTHOR'S CASES: CLINICAL RECORDS AND TABLES A—E.

Case 1. D.W., female, aged 14 years.

Family History. Father died of cerebral abscess, mother healthy; no history of nervous or muscular disease in any relation. Mother had 15 children, 9 of whom died, 6 being consecutive still-births; remainder are alive and well.

Past History. Last child of family, following 6 consecutive still-births; born at full time and healthy at birth. Sat up, talked, walked and cut her teeth at the usual times; no illnesses during infancy. Mother was 42 years of age at time of patient's birth.

On examination. Extremely thin in all parts except the face; intelligence and speech normal; can sit up and remain erect with ease, but walking is limited by deformities of the feet. The skin is pale, hair abundant but not coarse. There is a degree of wasting and atony of the musculature in general excepting the face; this is most marked in the limbs especially the hands. Loss of power is present but does not correspond in degree; ligaments are lax though hypotonicity is not extreme, no grotesque positions are possible. The fingers are very long, thin, and somewhat tapered, the ring and little digits are slightly contracted; no webbing, no clubbing nor acrocyanosis; nails are normal. Apart from the fingers the hands are narrow but do not appear unduly long; muscular wasting is marked both upon dorsal and palmar surfaces. Forearms and upper arms also appear long but as measurements show this is apparent rather than real, moreover their ratios are unaltered. Muscular wasting is marked and is also seen in the shoulder girdle.

Both feet clubbed and their relative length difficult to estimate; toes very thin and deformed, but no contractures present; no clubbing nor webbing, os calcis not spurred. Both lower legs and thighs are very thin, but not unduly long; all muscles wasted, especially glutei; there are no contractures; the external malleoli are prominent, patellæ of normal size but highly placed. The spine shows a slight dorsal kyphosis; both scapulæ are winged but of normal size and shape.

Radiograms of the hands suggest an increased length of phalanges and metacarpal bones which is confirmed by measurements, tapering is very slight and thinning not marked; no rarefaction and ossification normal. Feet show the curious dwarfing of the middle phalanges of the little toes peculiar to this case, the talipes causes foreshortening of metatarsal bones especially, their proximal extremities being very faintly outlined, in the right foot they are superimposed; thus accurate measurements are impossible.

The skull is dolichocephalic in shape, occipito-mental diameter is $9\frac{1}{2}$ in., greatest circumference $21\frac{1}{2}$ in. No asymmetry but jaw large and of the nutcracker type; fontanelles closed; supraorbital ridges normal; eyes not deeply set nor far apart; the palate narrow and somewhat arched; teeth normal; tongue not enlarged. Face not small but features are those of an adult; mouth and lips large; ears larger than normal, cartilage soft and deficient, lobules poorly differentiated, drums normal, hearing unimpaired. Radiogram of skull shows a marked reduction in size of the sella turcica and clinoid processes, with some thickening of the latter; no other abnormality. Chest somewhat flattened but otherwise normal, the rib epiphyses not enlarged. Lungs normal, no evidence of deformity of heart, viscera apparently unaffected, no defect in abdominal wall. Nervous system intact, all deep reflexes abolished but no true paralysis exists. Eyes normal, vision emmetropic, no dislocation of the lens, colour vision normal. No apparent affection of thyroid, thymus or other gland; external genital organs normal, early features of puberty present.

Urine normal; renal efficiency tests satisfactory; an estimation of the diastatic content of the urine showed that 1 c.cm. of urine contained 90 diastase units (discussed above). Fæces on a mixed diet showed complete digestion of all elements, especially no evidence of pancreatic deficiency; no abnormal pathogenic organisms found in cultures. Blood calcium content normal, amount of bilirubin present in the serum also normal; complete blood count satisfactory; a Sachs-Georgi reaction negative. Estimation of the basal metabolism showed a decrease in the rate of 30.6% compared with normal. Systolic and diastolic blood pressures low, but the latter was relatively high giving a small pulse pressure of 32 mm. Hg. Electrical reactions of

muscles showed a slight but equal diminution of response to both faradic and galvanic currents; the most atrophic muscles showed no reaction of degeneration, a moderately strong faradic stimulation caused much pain.

Case 2. K.S., female, aged 3 years.

Family History. Nothing of any interest to record.

Past History. Eighth child of a family of ten. Born at full time and healthy, though the long fingers were noticed; progress normal up to nine months when the amyotonic symptoms seem to have come on suddenly.

On examination. Thin generally; listless and utters no intelligent sounds but seems pleased with objects shown to her; quite unable to raise head from pillow; both arms are in full pronation, slightly abducted and flexed at the elbows; knees are flexed with some spasm; no contractures are present. no tremors, fibrillary twitchings nor choreiform movements. Symmetrical wasting of all muscles of both upper and lower limbs is marked, those of neck and shoulder girdle are slightly affected; in the face, trunk, and chest muscles normal. Loss of power is in proportion, in the limbs it is almost but not quite complete; in affected areas the recognition of muscle apart from adipose tissue is exceedingly difficult. Electrical reactions not tested. Hypotonicity marked but not extreme, limbs may be hyperextended without pain. The hands, especially the fingers, appear absurdly long, very thin and somewhat tapered towards the extremities; no webbing nor clubbing, nails are normal; muscles of the thenar and hypothenar eminences very wasted. The feet and toes are also of abnormal length, the calcaneus is markedly spurred; there is no tapering; muscles less affected than those of hands. Remaining parts of body do not appear unduly long and as measurements prove, their normal ratios are unaltered. Radiograms of hands show increased length, and measurements compared with the normal prove that this is present in every phalanx and metacarpal; moreover, as in other cases, this increase is out of proportion to the height; the terminal phalanx is the most effected; no rarefaction, ossification normal. Those of the feet do not suggest so marked an increase in length of the bones, accurate measurements are not possible; no tapering, no thinning, no rarefaction; ossification normal.

Skull normal in shape and size, fontanelles closed; the palate narrow and very highly arched; radiogram shows no abnormality. The face very small, features have an old appearance; ears have large lobules but otherwise normal; teeth are decayed but not malformed; skin and hair show no peculiarity. The chest is pigeon-breasted, there is a fibrotic condition at the base of the left lung; epiphyses not enlarged. Scapulae not winged, patellae normal, spine shows lumbar kyphosis only. Heart unaffected; abdomen prominent but no viscera felt. Nervous system intact, all deep reflexes abolished, but no paralysis present. Eyes normal in every respect. No apparent affection of thyroid, thymus or other gland; external genital organs normal.

This child developed an acute pneumonia and died before further investigations could be carried out. An autopsy was not performed.

Case 3. K.A., female, aged 5 years, 8 months.

Family History. Mother (now deceased) had very long and thin fingers from birth, and used to wear an unusually large size of boot; she was able to put her limbs into grotesque positions easily and without pain and she had a bilateral dislocation of the lens; she was 5 feet 8 inches in height and exceedingly thin; her weight is not known. She was the thirteenth child of the family and the only one affected, her father and mother were normal; she herself had four children of whom the two youngest were affected.

Past History. Youngest child of the family. Born at full time, healthy though the peculiarities of hands and feet were noticed at the time; except that speech was unintelligible until three years progress was normal.

On examination. Very thin and tall, speaks in a slow deliberate way but mentality is normal. Muscular wasting seen in palms of hands and to a less extent in soles of feet; in forearms and lower legs it is very slight and in upper arms and thighs it is absent. Scapulae not winged but seem long; slight lumbar kyphosis; remainder of the musculature is unaffected. Loss of power only in the hands very slight; marked hypotonicity present, ligaments are lax and extreme movements are possible without pain; both Achilles tendons slightly contracted

Hands appear very long, especially fingers which are thin and moderately tapered, slight but definite syndactyly present; no clubbing; nails are normal. No contractures or further deformities. Forearms and upper arms appear long and measurements show a marked increase above the normal, but ratios to the total body length are not disturbed, the increase is therefore in proportion to the height. Feet long and thin and toes slightly tapered; no clubbing; nails are normal. Os calcis spurred. Both feet plantar-flexed but full extension possible; no webbing; slight pes planus. Child takes size 13 in boots. Lower legs and thighs show the same proportionate increase in length as upper limbs; the external malleoli are prominent, patellæ are of normal size but highly placed.

Radiograms of hands show increased length, proved by measurements to be very marked and out of all proportion to the height, terminal phalanx is again most affected. There is some degree of tapering but no thinning, rarefaction absent and ossification normal. In feet this tapering is not evident, the bones appear to be increased in length.

Skull markedly dolichocephalic but symmetrical in shape; frontal bones, supraorbital ridges and lower jaw prominent, the palate narrow and highly arched, teeth normal, the tongue not enlarged. Face is old for age, lips very small, both ears very large with deficiency of cartilage and depressions in concha, crus of the helix badly defined, drums normal. Radiogram of skull shows a curious elongation of the sella turcica which is somewhat small in size, clinoid processes thickened but opening very little reduced in size. No other abnormality is seen. Chest normal, epiphyses not enlarged. Lungs normal, heart shows no deformity. Viscera unaffected. Nervous system intact; eyes show a bilateral dislocation of the lens with high degree of myopia, slight internal strabismus on the right side, colour vision is normal. No affection of thyroid, thymus or other gland; external genital organs normal.

Urine normal, also renal efficiency tests; diastatic content urine not raised in this case. Complete examination of stools satisfactory; blood counts normal, blood pressures low, Wassermann test not carried out; electrical reactions of muscles show no abnormality.

Case 4. O.A., male, aged 7½ years.

Family History. Brother of Case 3.

Past History. Third child of the family, born at full time and was healthy though the long hands and feet were noticed; speech was unintelligible until three years; otherwise progress normal.

On examination. Tall and thin, speaks in the same slow manner as sister but mentality normal. Apparent wasting of all muscles of forearms, hands, lower legs and feet, especially noticeable is atrophy of glutei; remainder of musculature unaffected, though there is slight lumbar kyphosis, and scapulæ which appear long have a tendency towards winging; both Achilles tendons slightly contracted. Loss of power in the hands, and slightly in forearms and lower legs; ligaments lax and hypotonicity marked, extreme movements can be carried out without discomfort. Hands, especially fingers, appear very long and thin; a slight degree of webbing, no tapering nor clubbing; nails normal, no contractures present. Forearms and upper arms longer than normal but in direct proportion to height. Feet long and thin but toes do not taper; no clubbing; nails are normal. Os calcis spurred, slight pes planus; takes size 1 in boots. Thighs and lower legs long but in proportion to height, external and internal malleoli prominent, patellæ of normal size but extremely highly placed. Radiograms of hands show same features as sister (Case 3) and measurements are actually comparable with those of a normal child of 14, i.e., 6½ years older. Radiograms of feet also similar to those of Case 3.

Skull also corresponds in every detail, the face old, the upper lip small and sunken and the abnormalities of ears are exactly similar. Radiogram shows sella turcica reduced in size with small opening, but no elongation as in the previous case; clinoid processes thickened. Chest somewhat funnel-shaped, with prominent lower sternum; epiphyses not enlarged; lungs normal, heart shows no deformity. Viscera unaffected. Nervous system intact. Eyes show bilateral dislocation of lens with high degree of myopia; slight internal strabismus on left side; colour vision normal. No affection of thyroid, thymus or other gland; external genital organs normal.

Urine normal; renal efficiency tests satisfactory; diastatic content of the urine not raised; examination of stools shows no defect; blood counts normal; blood pressures low but higher than in the previous case; Wassermann test not performed. Electrical reactions of muscles show no abnormality.

TABLE A.
SHOWING MEASUREMENTS OF LONG BONES IN INCHES WITH RATIOS TO HEIGHTS.

	CASE 1				CASE 2				CASE 3				CASE 4			
	Measurement		Ratio to height		Measure-ment		Ratio to height		Measure-ment		Ratio to height		Measurement		Ratio to height	
	Patient	Normal	Pat-tient	Normal	Pa-tient	Normal	Pa-tient	Normal	Pa-tient	Normal	Pa-tient	Normal	Patient	Normal	Pa-tient	Normal
	in.	in.	1 to :-	1 to :-	in.	in.	1 to :-	1 to :-	in.	in.	1 to :-	1 to :-	in.	in.	1 to :-	1 to :-
Total body length	53	57.5	—	—	32	36	—	—	47	39½	—	—	51	47½	—	—
Spine ...	24	25	2.2	2.3	15	17	2.1	2.1	19	16½	2.47	2.4	21	20	2.4	2.38
Upper arm ...	10½	12	5.0	4.8	5	5½	6.4	6.54	7½	6½	6.0	6.0	9	8½	5.7	5.75
Forearm ...	8½	9	6.2	6.4	4½	4½	7.5	7.36	7½	6	6.4	6.6	7½	7	6.7	6.8
Total arm ...	19	21	2.8	2.74	9½	10½	3.45	3.51	15	12½	3.1	3.1	16½	15½	3.1	3.1
Thigh ...	16	18	3.3	3.2	8½	9½	3.7	3.62	13½	11	3.5	3.6	14½	13½	3.5	3.5
Lower leg ...	12	15	4.41	3.83	7	8	4.6	4.5	12½	10	3.9	3.95	13½	12½	3.8	3.8
Total leg ...	28	33	1.9	1.7	15½	17½	2.06	2.05	25½	21	1.8	1.88	28	26	1.8	1.8
Foot ...	7½	9	7.0	6.4	5½	5½	5.56	6.85	8	6½	5.8	6.0	8½	7½	6.0	6.5
Weight ...	50 lb.	100 lb			16 lb.	30 lb.			42½ lb	42 lb.			51½ lb.	50½ lb		

TABLE B.
SHOWING THE LENGTHS OF PHALANGES AND METACARPALS IN MILLIMETRES OBTAINED FROM RADIOGRAMS (NORMAL FIGURES IN BRACKETS).

	Digit	Term. ph.	Mid. ph.	Prox. ph.	M'carpal	Sum total
Case 1	1st	14.0 (13.0)	20.0 (19.0)	34.0 (32.0)	52.0 (54.0)	120.0 (118.0)
	2nd	15.0 (14.0)	24.0 (23.0)	38.0 (37.0)	51.0 (52.0)	128.0 (126.0)
	3rd	15.0 (15.0)	23.0 (21.0)	36.0 (33.5)	45.5 (45.0)	119.5 (114.5)
	4th	12.0 (13.0)	16.0 (14.0)	27.0 (25.5)	42.0 (44.0)	97.0 (96.5)
	Thumb	20.0 (17.0)		25.5 (24.0)	36.0 (38.0)	81.5 (79.0)
Case 2	1st	9.0 (6.5)	11.5 (10.0)	22.0 (20.0)	29.5 (31.0)	72.0 (67.5)
	2nd	9.0 (7.5)	14.0 (13.0)	23.5 (23.0)	31.0 (29.0)	77.5 (72.5)
	3rd	9.0 (8.0)	13.5 (12.0)	23.0 (21.0)	28.0 (26.0)	73.5 (67.0)
	4th	8.0 (6.5)	10.0 (7.5)	18.5 (15.5)	23.5 (23.0)	60.0 (52.5)
	Thumb	12.0 (11.0)		16.0 (13.5)	21.0 (18.5)	49.0 (42.5)
Case 3	1st	13.5 (11.0)	18.5 (14.5)	32.0 (24.0)	51.0 (37.0)	115.0 (86.5)
	2nd	15.0 (10.0)	22.0 (17.5)	35.0 (27.0)	50.0 (36.0)	121.0 (90.5)
	3rd	15.0 (11.5)	21.0 (17.0)	32.0 (25.0)	44.0 (31.0)	112.0 (84.5)
	4th	12.0 (10.5)	14.5 (11.0)	24.5 (19.0)	40.0 (29.0)	91.0 (69.5)
	Thumb	17.5 (14.0)		23.0 (18.0)	36.0 (25.0)	76.5 (57.0)
Case 4	1st	14.5 (13.0)	19.0 (19.0)	32.5 (32.0)	52.0 (54.0)	118.0 (118.0)
	2nd	15.0 (14.0)	23.0 (23.0)	36.0 (37.0)	52.0 (52.0)	126.0 (126.0)
	3rd	16.0 (15.0)	22.0 (21.0)	33.0 (33.5)	46.0 (45.0)	117.0 (114.5)
	4th	14.0 (13.0)	16.5 (14.0)	26.0 (25.5)	42.0 (44.0)	98.5 (96.5)
	Thumb	20.0 (17.0)		25.0 (24.0)	35.5 (38.0)	80.5 (79.0)

TABLE C.

SHOWING THE RATIO OF THE SUM OF THE PHALANGES AND METACARPALS OF THE SECOND DIGIT TO TOTAL BODY LENGTHS.

Case	Ratio	Normal ratio
Case 1	1 to 10.04	1 to 11.4
Case 2	„ 10.03	„ 11.8
Case 3	„ 9.8	„ 10.1
Case 4	„ 10.2	„ 11.4

TABLE D.

SHOWING THE RATIOS OF THE SUM OF THE PHALANGES OF EACH DIGIT TO THE SUM OF THE PHALANGES PLUS CORRESPONDING METACARPAL BONES.

Case	1st digit	2nd digit	3rd digit	4th digit	Thumb
Case 1	1 to 1.76	1 to 1.66	1 to 1.61	1 to 1.76	1 to 1.79
Normal	1 to 1.84	1 to 1.71	1 to 1.66	1 to 1.83	1 to 1.92
Case 2	1 to 1.69	1 to 1.64	1 to 1.61	1 to 1.64	1 to 1.75
Normal	1 to 1.85	1 to 1.66	1 to 1.63	1 to 1.79	1 to 1.73
Case 3	1 to 1.8	1 to 1.68	1 to 1.64	1 to 1.8	1 to 1.88
Normal	1 to 1.74	1 to 1.65	1 to 1.58	1 to 1.71	1 to 1.78
Case 4	1 to 1.3	1 to 1.7	1 to 1.64	1 to 1.74	1 to 1.88
Normal	1 to 1.74	1 to 1.65	1 to 1.58	1 to 1.71	1 to 1.78

TABLE E.

SHOWING THE RATIO BETWEEN EACH PHALANX OF THE SECOND DIGIT TO THE CORRESPONDING METACARPAL.

Case	Terminal phalanx	Middle phalanx	Proximal phalanx
Case 1	1 to 3.40	1 to 2.12	1 to 1.30
Normal	1 to 3.70	1 to 2.26	1 to 1.40
Case 2	1 to 3.44	1 to 2.21	1 to 1.32
Normal	1 to 3.86	1 to 2.23	1 to 1.27
Case 3	1 to 3.33	1 to 2.22	1 to 1.43
Normal	1 to 3.60	1 to 2.09	1 to 1.33
Case 4	1 to 3.46	1 to 2.26	1 to 1.44
Normal	1 to 3.60	1 to 2.09	1 to 1.33