THE 'FROG' CHILD: A CONGENITAL LESION OF THE CORPUS STRIATUM?

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Out of 1,296 patients admitted to the Bath and Wessex Orthopaedic Hospital since its opening in 1926, four cases have come under observation which resemble each other, but which do not resemble any other cases admitted. So far as I can ascertain they do not exactly correspond to any clinical entity hitherto described in the literature, except for one case recorded by Fritsch of Breslau and quoted by Batten as a case of intrauterine poliomyelitis, though our present knowledge of this disease forbids us to accept the possibility of such a diagnosis. Fritsch describes the child in his case as having been born without injury through a breech presentation. The child had paralysis of both upper and lower extremities. There was a complete atrophic paralysis of both arms with loss of electrical excitability, while the legs were abducted to the fullest extent at the hips, and flexed at an acute angle at the knee so that the clubbed feet lay opposed to one another (cf. fig. 9). The muscles of the legs were not atrophic and responded normally to electricity. Batten remarks that the spasticity of the legs makes it unlikely that the diagnosis of poliomyelitis can be upheld.

Owing to the posture taken up by these children, which is similar to that of Fritsch's case, the Staff of the hospital has been accustomed to refer to them as 'Frog' babies, and they have been referred to under this term in a previous publication. This posture, which is the characteristic feature of the condition, consists in flexion and eversion of the limbs, more particularly the lower, maintained by a plastic rigidity of certain muscles similar to that of Parkinson's disease when this condition occurs without tremor (fig. 2).

PERSONAL CASES

The clinical notes of the four cases are as follows:—

Case I.—A.G. Male, born on March 12, 1925. Admitted to hospital on July 31, 1926, with a diagnosis of 'Contraction of knees in flexion and hips in flexion with external rotation; ? cause; also ? congenital absence of the patella.'

Family History.—Parents, three brothers and one sister alive and well. No prenatal injury or disease. Parturition normal, though labour was somewhat prolonged.

Previous Illness.—Double pneumonia, winter 1925.

Abnormality (curious posture of legs) was noticed from birth. This has not altered appreciably since. Before admission had prolonged massage and plasters
were applied to feet. The feet were improved but the hips and knees remained in statu quo.

State on Admission.—Certain rickety signs were present, viz. bulging forehead, wide fontanelle and slight Harrison's sulcus. The legs were in the posture described above (fig. 1).

Bones.—X-ray films showed bones to be normal with no rickety signs except that the upper ends of the shafts of the femora were rather expanded, with an S-shaped curve.

Joints.—The hips were held somewhat rigidly in flexion and external rotation and the knees in flexion; the right knee could only be extended to 135°. The right subastragaloid joint was in valgus. The left foot at first was in equinovarus but by the time of admission was almost normal.

Muscles.—The right quadriceps was very weak and the patella was at first thought to be absent, but later was found some distance above the knee joint. Both hip adductors were very weak. The invertors of the right foot were weak, but the opposing muscles were in a condition of plastic rigidity similar to that seen in striate lesions, and quite unlike anything hitherto observed in an ordinary case of rickets. This accounted for the peculiar frog-like attitude. The child was quite unable to walk or stand. There seemed to be no abnormality of the upper limbs or trunk at this time except for the aforementioned Harrison's sulcus.

Progress.—On August 17 the child was put on a Jones' frame with the right leg flexed, but with extension applied. By September 3 the right knee and hip could be straightened, but they returned to flexion when released. The left limb was by this time straight. On September 17 some limitation of extension of the elbow joint was noticed with muscular rigidity of muscles similar to that in the legs. On October 11 plasters were applied to try to correct the abnormal posture of the right leg and this treatment was continued with reapplications from time to time till February, 1927. In this month a piece of quadriceps muscle (which was rigid) was excised for examination and was found to be normal. A piece from the wasted biceps (which was weak and flaccid) was mostly fat and fibrous tissue. This muscle did not respond to the faradic current at the level at which the tissue was removed, although the whole
muscule did show some contraction. Manipulation was done while the child was under the anaesthetic, but no marked effect was obtained.

Later the boy suffered from whooping-cough and after recovery was fitted with a right calliper and at the end of June 1927 was also supplied with a left calliper and discharged home able to walk in these.

In spite of reasonably constant wearing of the apparatus, by May 1933 the condition of the right leg had relapsed to a considerable extent into its old 'frog' posture.

Condition on readmission, May 29, 1933.—The right hip cannot be extended beyond $160^\circ$ and cannot be internally rotated beyond the neutral position; all other movements are free. The left hip does not rotate internally beyond neutral, otherwise has full range. The right knee is contracted in flexion at about $140^\circ$; it has full active flexion. The right patella lies 3 in. above the line of the knee-joint. The right quadriceps will not keep knee even at full passive extension. The left knee extends and flexes completely. The right ankle is in slight equinus which can be overcome when knee is flexed. The left ankle comes just to a right angle. The right leg has $\frac{3}{4}$ in. shortening, probably due to flexion of knee and hip. The right calf has $\frac{1}{2}$ in. wasting; the right thigh 1½ in., 4 in. above knee. There is no fixed scoliosis. The child sits with winged scapulae, but this is not due to serratus weakness: rhomboids are fair. There is slight pigeon breast and Harrison’s sulcus. The patient is nearly nine, but is only about the size of a child of six. He is of reasonably good intelligence. He walks with a tilt of the body to the right. The right foot is in extreme valgus, the right knee pointing out; hip and knee are flexed. The left lower limb is now used in normal position except that the foot is in valgus. The right boot is badly worn down on its inner edge; the left has been thickened on its outer edge.

Traction and extension of the limb did not improve matters and tendon division by open operation was done. This gave a better result, but did not entirely correct the deformity. In September 1933 the child was discharged home with instructions to wear callipers by day.

Case II.—T.G. Male, born on June 12, 1925. Admitted on June 1, 1929, with a diagnosis of 'Frog' child.

Family History.—Father healthy. Mother has a goitre, but otherwise healthy. One brother 'delicate.'

Birth.—Labour was induced at eight months owing to narrowing of the maternal pelvis and because the mother was suffering from oedema.

Previous Illnesses.—Measles, chickenpox and pneumonia twice.

The abnormality was not noticed until the child ought to have begun to walk. He had always been weak but could not walk because of tightness of heels and flexion of knees and eversion of the hips. On admission at four years old he had not walked, but tried to shuffle about sitting on a chair. When he was two years old some 'spasm' of the fists was noticed by the mother. On admission he could feed himself clumsily; he could not write, but scribbled with his right hand.

State on Admission.—The skull was peculiar; there was marked asymmetry of the right frontal bone, while the left frontal was flattened; the left occipital bone was prominent as if the whole head had been squeezed obliquely. The chest was somewhat funnel-shaped with a definite Harrison’s sulcus. The fingers were slender. The spine was flaccid. The limbs were held in the posture of a squattting frog apparently by the peculiar postural tone of the muscles which showed a plastic rigidity suggestive of that met with in striate lesions.

X-ray films did not show rickety changes in bones; the epiphyses were normal.

Treatment by splinting, traction, and exercises in hot water met with only very partial success. Delay was caused by streptococcal infection of the throat; the child was always feeble and subject to infection. Later he was given a plaster bed and plasters were applied to his feet with some improvement. He later contracted whooping-
cough and returned home after recovery, but subsequently developed pneumonia and died. No autopsy was permitted.

Case III.—M.N. Female, born on September 23, 1930. Admitted on April 16, 1932, with a diagnosis of 'Frog' child.

*Family History.*—Parents healthy. One twin brother and three sisters healthy.

*Birth.*—Normal.

*Previous Illnesses.*—Asthmatic attacks since birth.

The disability was noticed when the child was 17 months old and thought to be bow legs. She had been able to 'walk' round holding on to chairs since the age of six months, but she could not yet walk unsupported.

*State on Admission.*—Head, chest and spine were normal. Upper limbs were normal except for a little thickening of the lower ends of the radii.

The hip joints showed a fixed external rotation which could only be overcome with difficulty. The knees showed fixed flexion at about 10° and the ankle joints slight calcaneus due to a plastic rigidity of the muscles reminiscent of that found in striate disease (fig. 2).

The only muscles actually weak were the glutei.

*Bones.*—The tibiae showed a little outward bowing and the hip coxa vara, but no subluxation. The epiphyses of each head were slightly irregular, but not such as would suggest any marked degree of rickets. The knee-joints appeared normal, with no widening of the epiphyses.

The 'frog' condition in this case was mild compared to the others, and as a result of plasters was much improved (fig. 3), but the rigidity was quite unlike that of an ordinary case of rickets. The child had difficulty in learning to walk, apparently preferring to go more sideways than forwards, although she eventually overcame this.

Case IV.—D.O. Male, born on March 21, 1924. Admitted on April 26, 1933, with diagnosis of 'Frog' child.

*Family History.*—Parents and one brother healthy. One died at three weeks from imperforate anus. One sister healthy.
Birth was normal, except for difficulty in breathing, the cord being wound twice round the neck.

Previous Illnesses.—Measles, whooping-cough, chickenpox and pneumonia three times. Tonsillectomy performed when aet. 4. Tracheotomy after measles.

The disability was noticed from birth as a weakness of all the muscles. He has never walked, but has crawled on hands and knees since he was about three years old. At this time his case was diagnosed as one of amyotonia congenita, but when he came under our observation he showed definite plastic rigidity of the muscles. He had been at a school for physical defectives and was of good intelligence.

State on Admission.—I.Q. 120 per cent. Incontinent of urine and faeces. There were marked bony deformities. The skull was flattened from before backward; the face was flat and very wide opposite the angles of the lower jaw (fig. 8). The spine showed a long right thoracic scoliosis with much distortion of all the ribs which were prominent posteriorly on the right side (figs. 6 and 7). The chest was deep and funnel-shaped with much spreading of the ribs in front. There was some bowing of the upper third of each tibia. The os calcis was almost vertical and the cuboid had a spine on the dorsum. There was extreme plantar flexion of the big toes at the metatarsophalangeal joints and to a less extent of the other toes, which also overlapped one another (fig. 9).

The tendon reflexes were not obtained. The posture lying down was with each hip in extreme external rotation and slight abduction. The knees were flexed at 160°. The feet were in equinus, the toes flexed with the outer border of each foot on the bed. This posture was maintained by plastic rigidity of the muscles.

X-ray films showed no characteristic rickety changes in the bones in spite of the marked deformity (figs. 4 and 5).

While the child was in hospital control of organic reflexes was established and by persevering with orthopaedic measures he learnt to walk.
FIG. 6.—Case IV. Shows scoliosis.

FIG. 7.—Case IV. X-ray photograph of spine.

FIG. 8.—Case IV. Shows peculiar shape of skull and facial bones.

FIG. 9.—Case IV. Shows deformity of feet.

DISCUSSION

At first sight these four cases (three males and one female not related and with no history of a similar condition being noted in the family of any of them) might have been taken for somewhat unusual cases of rickets, and they undoubtedly presented certain features of this disease. The children were
all somewhat delicate in their physical constitution but of average or good intelligence. Of course, it is quite possible that the true diagnosis should be simply one of rickets, but against this they presented a characteristic posture of the limbs of flexion and external rotation which is not commonly met with as a rickety deformity. The X-ray films disclosed no epiphyseal irregularity, such as would have been expected had the extreme degree of deformity been due to rickety changes. No benefit was obtained even in the infantile case by administration of vitamin D and exposure to sunlight. Above all, the rigidity of the muscles was quite unlike anything seen in ordinary cases of rickets, in which the muscles are atonic and weak. Other gross bone and joint disease was excluded by the X-ray appearances.

The trouble therefore would appear to be in the neuromuscular apparatus and not in the bones or joints.

The condition does not in any sense resemble a diplegia or double hemiplegia since the posture is the reverse of the adducted extended limb familiar in these conditions, though it is not impossible that some hitherto undescribed cortical lesion might produce a condition the opposite to diplegia. However, as has been stated, the rigidity is plastic rather than spastic. There is no hyperkinesis such as is seen in Vogt's double athetosis or Wilson's disease, and the group of previously described lesions of the corpus striatum.

The condition of myodystrophia fœtalis deformans or arthrogryposis multiplex congenita must be considered, especially in connexion with Case IV, in which the feet might be reminiscent of this. They were not, however, clubbed, but in an exaggerated calcaneus position.

Stewart Middleton has recently given an excellent survey of this affection. For nearly a century orthopaedists have recognized a well-defined congenital deformity affecting both upper and/or lower extremities. The deformity is present at birth and consists of club-hands and club-feet without any bony aplasia to account for its presence. It is accompanied by more or less limitation of the normal range of movement at the joints of the affected limbs. The mobility of the joints may be only slightly curtailed or the limbs may be, in the more severe cases, almost fixed in any degree of flexion or extension.'

Where passive movement of the joints is possible, and this is by no means in every case, it is found that active movement is either absent or is greatly diminished by a muscular weakness which is explained by the observation that all the muscles involved are much wasted. There results a relative prominence of the joints which has given rise to the impression that the rigidity of the limbs is due to a primary articular lesion. Where the lower extremities are affected, congenital dislocation of the hip joints is frequently present and in the more common cases, where the knee joints are fixed in hyperextension, congenital dislocation of the knees with absence of the patellæ may result.

In the present series of cases the posture is different, for in myodystrophia
there is a tendency for hyperextension and the club hands and feet are characteristic, while in these ‘frog’ children the eversion and flexion of the hips is in no case associated with congenital dislocation of that joint.

The muscles are notably wasted in myodystrophy, but this was not the case in the present series except in the first. Again, the affected muscles which are responsible for the deformity show characteristic microscopic changes. They were found to be composed very largely of fat. Amongst the fat could be seen occasional oval cells with central nuclei. They were clearly muscular in type and might be interpreted as undeveloped muscle-cells in the myoblastic stage of differentiation, or as a remnant of degenerating or degenerated myocytes. In other muscles were found varying proportions of normal muscle fibres in a stroma of fat. As has been said already, in myodystrophy the wasted fibro-fatty muscles produce the deformity, whereas in the case described above the muscle causing the deformity was normal, while the opposing weak muscle was fibro-fatty. The conditions, therefore, seem not to be parallel. In myodystrophy the contracture is completely resistant, while in the ‘frog’ children a considerable improvement was obtained by persistent stretching and re-education.

It is suggested that these four cases represent a clinical entity which can best be explained by some congenital lesion of the corpus striatum or other part of the extrapyramidal arc, causing the characteristic plastic rigidity without hyperkinesis which produces a deformity of the limbs, more particularly of the lower, in a position of external rotation and flexion. The condition is associated, especially in Cases II and IV, with marked skeletal deformities not accompanied by characteristic bone or joint X-ray appearances. The distortion of the skull and spinal column in these two cases is not, however, accounted for, and the absence of post-mortem examination makes the whole presentation of these cases highly speculative and of uncertain value. It is possible that others who have met with similar cases may have been more fortunate in elucidating the cause of the condition and so be led to throw light on an obscure problem.

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