Remarkable congenital deformity, partly resulting from constriction of the umbilical cord. Presented to the Pathological Society. Session 1847-48 / [William John Little].

REMARKABLE

CONGENITAL DEFORMITY,

PARTLY RESULTING FROM CONSTRICTION BY THE UMBILICAL CORD.

PRESENTED TO THE PATHOLOGICAL SOCIETY,

SESSION 1847-8.

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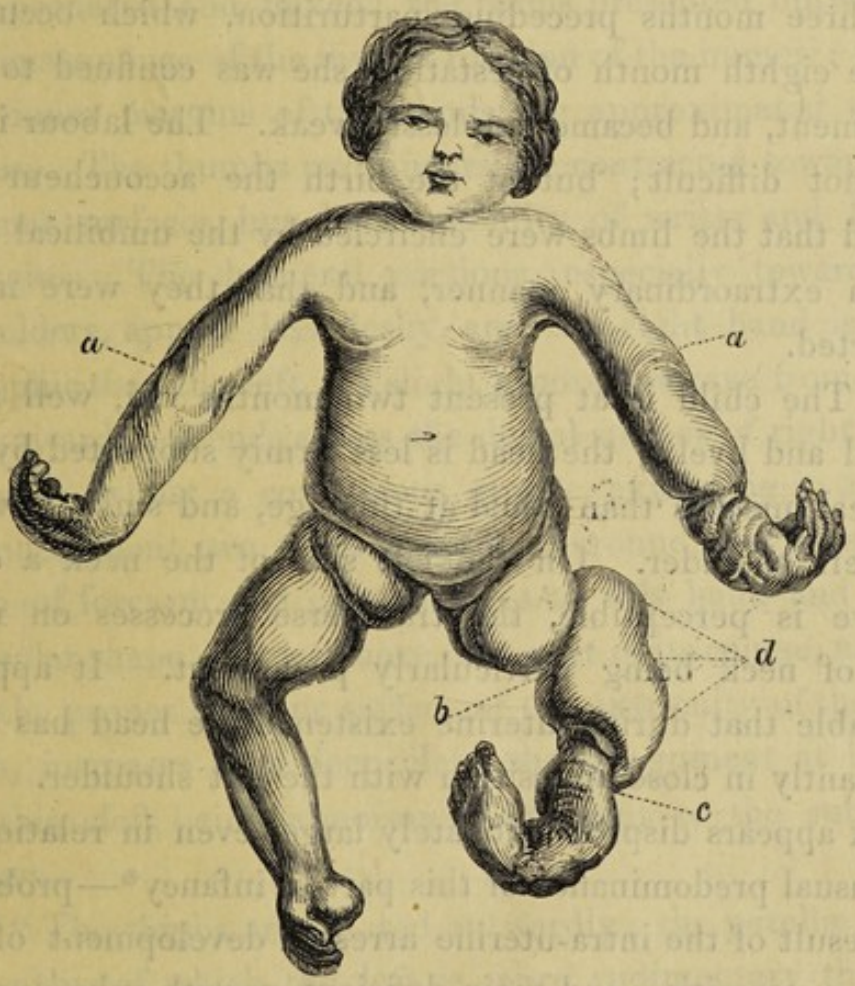
REMARKABLE CASE

OF

CONGENITAL DEFORMITY.



The following instance of congenital malformation has appeared to me sufficiently interesting to merit the attention of the Pathological Society. The case appears as follows in my journal:

"*August*, 1846. — A male child brought under my

notice: — the father a person of literary and sedentary habits, the mother a person of delicate frame, subject to dyspepsia, hepatic derangement, hemorrhoids, attacks of constipation lasting from ten to twenty days — the bowels, in fact, rarely acting except at intervals of several days; she has borne one other child, at present aged about three years, delicate, and liable to "fits" during dentition. Whilst pregnant with the subject of these remarks, she suffered greatly from the movements of the fœtus, and from pain in the right iliac fossa. During three months preceding parturition, which occurred at the eighth month of gestation, she was confined to her apartment, and became helplessly weak. The labour itself was not difficult; but at the birth the accoucheur observed that the limbs were encircled by the umbilical cord in an extraordinary manner, and that they were much distorted.

"The child is at present two months old, well nourished and lively; the head is less firmly supported by the cervical muscles than usual at this age, and sinks towards the left shoulder. On the left side of the neck a deep groove is perceptible, the transverse processes on right side of neck being particularly prominent. It appears probable that during uterine existence the head has been constantly in close apposition with the left shoulder. The trunk appears disproportionately large, even in relation to the usual predominance of this part in infancy*—probably the result of the intra-uterine arrested development of the lower extremities, and their deficient growth subsequently to birth. The four extremities are malformed, each of the upper extremities being similarly affected, whilst in the lower limbs the deformity appears to differ on each side. Examined in detail, the arms are observed to be

* The drawing is in this respect inaccurate.

inwardly rotated to the utmost extent, the shoulders prominent from deficient development of deltoides; superior articulations of the humeri movable, but the limbs can be carried upwards and outwardly to a less extent than in other directions. The olecranon processes present outwardly and somewhat anteriorly; very slight mobility at elbow-joints exists; in consequence of the inward rotation of humeri, the forearms are moved during flexion towards the dorsal, rather than towards the anterior aspect of the body, as in the natural formation of these organs. The wrists are pronated and flexed; the palms presented outwardly, in consequence of the inward rotation of the humeri; whilst the inner margins of the hands are approximated to the ulnas. The thumbs and fingers are contracted towards the palmar surfaces, but little mobility of wrists and fingers remains. The humeral portions, especially towards the shoulders, appear less fleshy, and the right hand is more atrophic than the left. A slight groove, concave from above downwards, extends across the dorsal surface of right metacarpus; whilst a continuous groove, extending in a spiral manner about two turns and a half around the lower portion of forearm and carpus, gives to this hand and arm a singular shape. This groove, in a part of its course, namely, on the proper anterior surface of the extremity of the forearm, increases to a deep cleft, the integument at bottom of this cleft being abnormally adherent to the subjacent bones.

"The thighs are rotated outwardly; the patellæ consequently, of which the left is more rudimentary than the right, present externally; their situation is marked by a deeply wrinkled depression in the integuments, to which they appear to adhere. The knees are somewhat contracted, the obstacle to their extension being firm, the left more unyielding than the right. The feet have the form

of *talipes varus*, the right assuming the intermediate, and the left the most complete grade of that affection. A crease, deeper on the internal than on the external aspect, extends spirally around the middle of the right thigh; whilst four spiral turns of a deep cleft exist in the left limb, two above and two below the knee-joint. About the middle portions of the left thigh and leg respectively, two spiral turns (*b* and *c*) constitute fissures so deep that the cutis is here in close proximity to the bones; the intermediate portion of the member (*d*) being connected with upper portion of the thigh, and the lower portion of the leg, by the attenuated thigh and leg bones, invested in these situations by very fine integument only. In fact, where the two principal clefts occur, the distal portions of the member are maintained in connexion with the proximal portions by merely the shafts of the bones, the nerves and bloodvessels, and thin laminæ of cutaneous tissues. The parts of the member placed between the two principal grooves are, on the contrary, bulky—preternaturally developed."

Such were the deformities in this unfortunate infant, whose functions of organic life were naturally performed. My opinion being requested respecting the practicability and propriety of adopting remedial means, I was induced (as the result of my observation in other, in some respects similar, cases) to state my belief that, whilst the lower extremities might be sufficiently restored for the purposes of imperfect locomotion, and the upper extremities improved to the extent of enabling the infant to approximate the hands tothe mouth, yet that very slight general function of the hands and arms would be obtained. Having represented to the parents, who were desirous of relieving as far as possible the infirmity of their child, that the period of infancy afforded greater facility for restoration

of flexibility, and consequent proportionate use of the limbs, than more advanced age, it was determined at once to commence the treatment. Section of Achilles tendon in both feet, that of tibialis posterior tendon in left foot, application of splints, with manipulations to the ankles, knees, and wrists, and a spring instrument applied for gradual flexion of the elbows and inward rotation of humeri, sufficed, within six months, to effect first part of treatment, namely, the restoration of nearly complete form and flexibility. Three months later, the child had acquired some power; he was enabled to shrug the shoulders, inwardly rotate arms, and grasp toys with the hands, but had obtained no capability of voluntarily acting upon the deltoids and raising the arms; the voluntary movement of the elbows was exceedingly limited.

The form and flexibility of the feet were fairly restored; the processes of the left tibio-tarsal articulation remained sparely developed; the knees were much straighter, and more movable; the outward rotation of the thighs much less constant; the child could playfully throw these limbs about, adduct them, partially bend the knees, and gave promise of realisation of the expectations entertained of their restoration to a certain degree of usefulness.

At eighteen months, however, the child, having occasionally suffered whilst cutting teeth, was carried off somewhat suddenly by convulsions.

Death having occurred at the distance of 200 miles from London, the opportunity of post-mortem investigation was not afforded me.

I now proceed to suggest to the Society what appear to be the pathogenetical and pathological points of principal interest:—

1. The influence of the mother's state of health prior to and during gestation.

2. The question of the dependence of the deformities upon derangements within the fœtal organism; or,

3. Their production from the influence of causes acting externally upon the fœtus.

4. The probability of the umbilical cord having been the influential external agent.

5. The illustration afforded by this case of the manner in which certain uterine arrests of development take place: for example, spontaneous amputation in utero.

6. The probability of such restoration of functions as would have rendered the individual capable of administering to his natural wants, viz. assumption of food, locomotion, &c. being obtained; and therefore of the child being rendered independent of external aid for the bare support of life.

Experience, in my opinion, abundantly testifies, that illness on the part of the female parent, whether it operate by directly lowering the tone of the nervous system, or by abstraction of the nutritive materials of her frame, constitutes one of the most powerful of the predisposing causes of congenital deformity. In the instance under consideration, an impaired nervous system and ill-nourished frame of the mother existed; and, in addition, the fœtus was exposed to other injurious influences consequent upon the mother's compulsory total avoidance of exercise during later months of gestation, and the habitual inordinate confinement of the bowels. So inveterate was the constipation, that her only mode of defecation consisted of the evacuation of a few scybalæ at intervals varying from fourteen to twenty-one days. The inability to use bodily exercise was unquestionably prejudicial; and it appears not very improbable that the constant accumulation of fæces in the colon and rectum, indurated as these excrementitious matters become by long residence in the interior of the

body, small portions only being discharged at intervals of many days, may operate in the manner of a solid tumour lodged in the pelvic and abdominal cavities, and occasion disturbance in the functions of the gravid uterus and of its contents.

Whether or no constipation may operate in the mechanical manner here glanced at, it cannot be doubted that habitual constipation, by disturbing functions of the alimentary tube of the parent, may have contributed to the production of the infant's deformity.

The origin of the deformities from derangements occurring within the fœtal organism, is a proposition the proof of which is more difficult. This particular instance of deformity is not referable to primitive imperfection of the germ—unless it be supposed that an hereditary predisposition to deformity existed, of which, however, no evidence could be elicited—no excess or deficiency of parts was observed, that might bring the case within the category of malformations originating from that cause.

Elsewhere* I have advocated the dependence of numerous distortions upon derangements of the cerebro-spinal system of the fœtus, induced by a variety of causes. The inquiry whether this solution is applicable in the present instance will be facilitated by a reference to the anatomical pathology of the parts principally affected. The upper extremities appear symmetrically affected; inward rotation of arms — extension of elbows — extreme pronation, with flexion of wrists and fingers — deficient development, with apparently entire paralysis of deltoids — apparent degradation of elbow-joints, possibly the result of excessive persistent action of the pronator muscles, which were much

* *Symbolæ ad Talip. varum cognos.* Berlini, 1837.

"Treatise on Club-foot and Analogous Distortions." Longman and Co.

shortened and tense—contraction of flexors of wrists and fingers—wasting of palmar tissues, the thumbs being especially atrophied and drawn into the palms—apparent paralysis of extensors of wrists and fingers:—such were the numerous lesions of these important members.

The deformity of the lower extremities is more symmetrical than a superficial examination might suggest—the knees being flexed and contracted, the feet deformed as in *talipes varus*—the commonest form of congenital deformity. This symmetry in the affection of both lateral halves of the body, combined with existence of contraction of certain muscles (the most part flexors), and paralysis of other muscles (extensors), gives strength to the supposition that a cause acting through the medium of the cerebro-spinal system of the embryo occasioned the deformity.

But whilst disposed to attribute the distortion of the various articulations to causes operating through the medium of the nervous centres, certain changes in the form of the limbs, as the grooves and deep clefts already described, irresistibly lead to the conviction that agency external to the fœtal organism had been influential in augmenting the departure from the normal form; and the observer promptly accuses the umbilical cord as the instrument by which the fissures and intervening protuberant parts of the limbs were effected. The smooth, pulley-like appearance of the grooves upon the left hand and foot present a width proportioned to the size of the funis. No stretch of the imagination is required for the perception of a spiral tendency in the fissures—a sufficient evidence that they were produced by a single cord convoluted about the parts, and not by a series of separate bridles. The enlargement of those parts of the lower extremities situated below the fissure on each thigh contiguous to the pelvis, and of the parts between the fissures lower down

on the left thigh and leg, was doubtless produced by the impediment offered at the fissures to the return of blood by the veins. In those parts of the members where the constriction of the cord was only moderate, a groove, proportioned in width to the thickness of the funis, resulted from the compression; whereas the comparative narrowness of the deep clefts probably resulted from the reduction in thickness of the funis, consequent upon the greater degree of tightness with which it encircled the limbs in these situations. The constriction exercised upon the left limb had, in the middle part of the thigh and about the middle of the leg, been so considerable, that in both of these situations the whole of the tissues of the limb, excepting the bone and certain bloodvessels and nerves, with exceedingly thin layers of velvety integuments, had been severed. As already remarked, the circulation and consequent vitality of the distal part of the member had been maintained by the minute vessels and nerves contained in the unsevered tissues. Apparently, a breach of surface had never existed, pressure having effected the division of tissues through the agency of interstitial absorption. It can scarcely be doubted that, if the entanglement and constriction of the limbs by the funis had taken place at a very early period of fœtal existence, and had been sufficiently complete to have produced strangulation of the distal part of the members, entire separation, amputation of the limb in utero, would have been determined. The child in that case would have been born apodic; a stump only of the former limb, furnished with a nipple-shaped process at the extremity, would have remained, such as I have observed to exist in the stump of spontaneous amputation, and such as surgeons are but too familiar with after ordinary chirurgical ablation of members.

I may be permitted to dismiss the subject of treatment with the remark, that the result of the means employed fully justified the prognosis recorded. If the life of the individual had been prolonged to the age of seven or eight years, it is probable that the various contractions would have finally been overcome; that the debilitated muscles would have acquired the capability of responding to volition; that the form of the articular surfaces—doubtless in some degree modified by malposition—would have been restored to a form consistent with usefulness. The imprints of the pressure of the funis would have permanently remained, although they might not have rendered the muscles interested at the strictured parts entirely void of functional activity.

I shall conclude these observations upon this case by adding my belief that the links in the chain of pathogenetical causes were as follow:—Impaired constitutional power in the parent having predisposed to derangement in the cerebro-spinal system of the embryo—intra-uterine convulsions of the fœtus—atony and paralysis of certain muscles, with inordinate action of other muscles, as consequences of such cerebro-spinal derangement—entanglement of limbs by funis (itself the accidental result of the convulsions, but the cause of the uncommon spiral grooves, fissures, and clefts). I am not unaware of the possibility that the whole of the deformity may appear to have been produced by mechanical causes; but offer my opinion, derived from comparison with, and analogy in, other cases, that in this instance the singular coincidence existed, of causes of deformity operating within the fœtal organism concurrently with the action of other causes externally influencing the development of the members.

The popular idea, of dependence of congenital de-

formities in general upon the imagination of the female parent, is as much the ultra-dynamic view of the origin of distortions, as the unphilosophical opinion of medical writers in by-gone periods, advocated by some surgeons almost to the present day, of the production of club-foot and similar deformities by malposition in utero, is the ultra-mechanical view of this interesting subject.

An extensive view of the pathogeny of congenital distortions and malformations in general, proves, in my opinion indisputably, that, although individual instances of deformity may, from the probable nature of the first genetic impulse, be referred either to the extreme dynamic, or to the mechanical theory of the origin of deformities, the majority of distortions and malformations spring individually from causes that may be arranged in a category removed as much from the popular idea of maternal imagination on the one hand, as from mechanical influences on the other. Indeed, a sound pathology of the majority of deformities may occupy a position intermediate between these extreme views.

The result of the study of the pathology of deformities congenital and acquired, is the approximation of these apparently different affections to each other, and to pathology in general. Almost every congenital distortion has its analogue in an acquired deformity, precisely as many diseases of extra-uterine life have their representatives during fœtal existence.

The belief in the agency of the umbilical cord as a cause of congenital deformity has been very generally entertained;* yet I am not aware of any recorded examples of the kind in fœtuses that have survived birth. In acephalic monsters, traces of constriction by funis are often perceptible. Elbin (*Dissertatio de Acephalis*, Berlini,

* Sömmering, v. Baue d. menschl. Körpers. viii. 1. s. 454.