CONGENITAL KYPHOSIS.

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The congenital kyphosis described in the following case was found in association with a cervical rib and a congenital high scapula. Cervical ribs are not uncommon, congenital high scapula is by no means a rarity, and though there is not necessarily an association between these two, it is, I think, not unlikely that in certain cases of high scapula the malposition is due to peculiarities of development of ribs or spine. I would class such cases of high scapula in a special division as one classes cases where osseous union exists between the scapula and the spine.

In the Edinburgh Medical Journal (September 1910) I published a case of congenital scoliosis due to the inter-position in the spinal column of an extra half vertebra. The literature of this subject is not very extensive, and, except in post-mortem examinations of foetal abnormalities, there are not many cases of this condition described. It is, however, one which can be readily understood, for peculiarities of segmentation are not rare, the inter-position of a whole vertebra is not uncommon, and the suppression of a whole vertebra is known to occur; and it is obvious that the inter-position of a half vertebra or the want of development of half a vertebra would produce a congenital lateral curvature. While reflecting on the production of congenital scoliosis previous to publishing the case, it occurred to me as possible that other defects in the development of the vertebrae might produce even more uncommon spinal deformities. Defects in the development of the neural arches explain the osseous abnormalities of spina bifida, but defects in the development of the bodies of the vertebrae must produce lateral or angular curvature. What these deformities might be can be premised from a consideration of the development of the bones in question. According to Cunningham the ossific centres for the bodies first appear in the lower thoracic vertebra about the tenth week. According to Keith the ossification of the centra begins in the 6th dorsal vertebra at the seventh week of embryo life and proceeds towards the extremities of the spinal column from there, and the ossification of the neural arches begins at the same time, but in the 1st cervical, and proceeds downwards towards the
cauda. "An oval nucleus develops in each body. At first it is placed dorsal to the notochord, but subsequently surrounds and causes the disappearance of that structure. Occasionally, however, the primitive centre appears to be formed by a coalescence of two primary nuclei. Support is given to this view by the occasional occurrence of vertebrae in which the body is developed in two lateral halves or in cases where only one half of the body persists" (Turner). We do not need to seek the explanation of the suppression or inter-position of a whole vertebra leading to a shortening or lengthening of the spine with retention of normal axis, but the paragraph just quoted offers the explanation of the suppression or inter-position of a half vertebra leading to the production of a congenital scoliosis. Consideration of the development of the embryo also explains an abnormality of far rarer occurrence, namely, congenital kyphosis. There is no reason to cavil at Cunningham's statement that the body of the vertebra is developed from an oval nucleus situated, in its first appearance, dorsal to the notochord; and it seems conceivable that the ossification of the peri-notochordal cartilage might not take place in the usual way and to the usual extent. If, then, an abnormality did occur, it would not be in the posterior part of the body of the vertebra, where the ossific centre originally appears, but in the anterior part to which ossification extends later. What would happen in such a case is obvious, viz., that the oval nucleus commencing posterior to the notochord would ensure the posterior part of the body of the vertebra being ossified first, and if the ossification did not extend forwards the centrum would be unequally developed antero-posteriorly and one would get a kyphosis. Embryologists do not mention the possibility of a primary centre of ossification appearing in the cartilage anterior to the notochord. The failure of the ossification to extend anteriorly would produce an exact reproduction of the collapse of a vertebral body as caused by disease. It has been pointed out by Ballantyne 7 that disease in the bodies of the vertebrae in the foetus is not unknown, but it must be in the immense majority of cases merely one manifestation of a much more general affection, and there can be no difficulty in pathological differentiation between a disease and a developmental defect. In looking through the literature of congenital scoliosis and defects of the spine I have not found congenital kyphosis hitherto described, but a case which cannot be otherwise explained was, some three years ago, sent to me by Dr. Burgess of Stanley.
Fig. 1.—Congenital Kyphosis showing the High Scapula.
Fig. 2.—Radiogram of Congenital Kyphosis, showing the Presence of a Left Cervical Rib.
The patient was a boy of two years and six months of age. He was the youngest of three; one child had been found dead in bed when ten weeks old, the other child and the parents were well and healthy. The patient was a full-time child, breast fed, walked at fifteen months, cut two lower teeth at six months, but was always a fretful child and required a good deal of nursing. It appeared, however, that he had been always quite strong, and he had no record of illness. When the child was six months old the mother noticed a swelling in the back at the root of the neck and towards the left shoulder, and she thinks it got more obvious as growth proceeded. When he came under my notice in January 1913 the child presented a healthy appearance. He had a fair complexion, wide open blue eyes, pouting lips, and a curiously well-marked droop of the cheeks, reminding one strongly of the appearance of a "Brownie" in a child's picture book. The child walked well, and the deformity could not be observed until he was undressed. He was well nourished, the skin was in good condition, but the child was rather small for his age, though in this relation it must be noted that both parents were under the average stature. The front view of the chest presented nothing abnormal, the skull was asymmetrical, the left parietal angled more from the sagittal suture, and the right parietal eminence was the more distinct. There was an obvious projection in the upper dorsal spine, and as he sat with his hands symmetrically placed in front of him it was noted that the inferior angle of the left scapula was much more prominent than the right and was placed an inch and a quarter higher on the chest. The spinous processes of the vertebrae appeared to be accurately superimposed, but when one approached the top of the dorsal region there was a prominence about the last cervical spine which was accentuated by what appeared to be a forward displacement of the fifth or sixth cervical vertebra. The left scapula was placed higher on the chest than the right and projected slightly into the neck. It lay rather more obliquely than normal and was not so large as its fellow. There was no abnormal connection osseous or cartilaginous, between the scapula and the spine. The movements of the scapula and the shoulders were quite free and full, but apparently from the obliquity and small size of the scapula the supraspinatus and infraspinatus muscles had not the normal purchase on the humerus.

The parts were radiographed by Dr. Pirie, but of course the details in the shadow cast by the spine are obscured by the shadows of the sternum and clavicles. The high position of the left scapula is obvious. It is associated with an irregularity of the vertebral bodies and of the ribs. As regards the latter there appears to be a cervical rib, and there may be some fusion of the 2nd and 3rd ribs. The third intercostal space in its posterior part is unduly wide as compared with
the right. What has happened to the vertebral bodies is more difficult to make out, for no radiogram shows them clearly at the affected part. With the help of the fluorescent screen, however, and examining the parts in various obliquities it appears that the abnormality is in the 1st dorsal or 7th cervical vertebra, and consists in a defective development of the anterior part of the body of either of these two vertebrae. The radiogram seems to show a slight tilting forwards of the upper three dorsal vertebrae as if the abnormal development had affected their vertical axis, but this is entirely overshadowed by the angular curvature at the junction of the dorsal with the cervical spine. It is possible that the development of the anterior segments of the bodies of the immediate subjacent vertebrae may have been secondarily affected by the alteration in the axis of transmission of the weight above the 1st dorsal on account of the bony defect.

One can scarcely include in the same category as the above case "A Case of Congenital Kyphosis" reported by Dr. L. Bernhard, though the title seemed likely to throw some light on the condition. The child was 39 days old, the third child of a healthy family, though the eldest had died "in consequence of abnormal birth." There had been no trouble at the birth of this child, but it "had not cried since birth." There was a kyphosis in the upper part of the dorsal spine extending from the 2nd to the 7th dorsal vertebra, comparatively regularly arched, which could only be slightly straightened by laying the child flat. Bernhard saw the child again six months later when, apparently, defective mental development was obvious, and for a month the child had had recurring twitchings of the trunk "as if the child were bowing." The growth of the skull was defective and there was alleged to be a total synostosis of the skull sutures and the fontanelle. The kyphosis "which was so evident on the first examination is no longer to be observed," but a slight left scoliosis had developed in the mid-dorsal region. Bernhard, in discussing this part of his case, says, "the disappearance of the kyphosis in spite of its earlier relative strength is interesting. We may assume that the lengthy opisthotonic straightening of the back bone had operated on the kyphosis so as to bring about its obliteration and the softened vertebrae had taken on a slight lateral displacement."

A regular dorsal curvature occurring in a congenital idiot, and this curvature disappearing within six months, is surely not to be classed with the case I have just related.
I have in my museum two skulls of congenital idiots the subjects of premature synostosis. In both these children a postural kyphosis of the spine existed, and the restriction of movement in the spine seemed to me to be part of the diplegia with contracture with which each was affected. The kyphosis, as in Bernhard’s case, was merely a secondary accompaniment of an extensive neurological congenital defect.

Though I know of no further literature on congenital kyphosis, there is an analogous condition described in the Medical Record in relation to the skull, under the term “Basilar Kyphosis: its Relation to Certain Cerebral Deformities,” by S. E. Post.6 “By basilar kyphosis is meant a deformity of the skull homologous with kyphosis in the spine.” It is pointed out that though the prominent angle of spinal kyphosis is obviously the spinous processes, the essential deformity lies in the bodies of the vertebrae; and though the possibility of congenital spinal kyphosis is not suggested, the author’s observations are made in relation to certain deformities of the skull depending on congenital defect in the development of the basilar process of the occipital bone, the post-sphenoid and the pre-sphenoid. These osseous elements form the os tribasilaire of the skull-base and are homologous with the bodies of the spinal vertebrae, as Virchow originally pointed out. The pre-sphenoid and post-sphenoid bones are ankylosed before birth and constitute the body of the sphenoid, but the occipital and sphenoid do not fuse until the 18th or 20th year, and it is between the sphenoid and occipital (the occipito-sphenoidal articulation) that the deformity or alteration of relationship usually occurs. “Basilar kyphosis is not simply a persistent foetal trait, it is a distinctly abnormal or pathological condition” the author states, and adds that Ackerman found it in a number of cases of encephalocele.

The main point made by the author in that paper is that the deformity bears a causal relation to certain defects produced by it in the cranial capacity and so on the brain. In congenital spinal kyphosis the symptoms need not be those of pressure on the contents of the neural canal, but it is interesting in my case that the deformity was in association with defects in ribs, and a malposition of the scapula.

In a previous number of the Edinburgh Medical Journal5 I described cases of congenital high scapula which had come under my notice, and I emphasised that such a congenital defect is not uncommonly accompanied by other congenital defects. Thus,
there may be spina bifida (vera or occulta), and in one of my cases there was suppression of ribs, but I know of no case published where congenital high scapula has been in association with an angular curvature of the spine. The defects in the spine and the ribs are defects of osseous formation—the defect in the scapula is not a structural one, but a malposition followed secondarily by improper growth in accordance with ordinary physical laws. I possess the skeleton of an extreme case of fusion of ribs, and this fusion is in association with remarkable irregularity of segmentation in the spinal column, several centra being partially suppressed.

The question arises, Is the defect a primary congenital one of the scapular position or one of the vertebrae? The answer to this seems to lie in the chronological sequence of events in the ossification of the bones involved.

I have already stated that, according to Keith, ossification in the bodies of the vertebrae commences in the 6th dorsal at the seventh week and proceeds upwards and downwards from that bone until the ossification of the axis takes place in the fourth month. Should any abnormality occur, it would only be in accordance with what happens elsewhere to find it take place at the transition from the dorsal to cervical vertebrae or dorsal to lumbar. On the other hand the scapula is, in the human embryo, originally cervical, and descends during the second month, to reach its final position in the third month. The change in position of the scapula is intimately associated with the elongation of the cervical region, and is accompanied by such alterations in the muscles and nerves that it is likely more than one factor may prevent the scapula's descent. It is nothing that the primary centre of ossification for the body of the scapula appears in the second month of foetal life, for in high scapula we have to deal not so much with when its ossification commences as with when its translation from the neck to the thorax takes place. It seems credible at any rate that interference with the ossification of the vertebrae, which commences at the seventh week, would be more likely to interfere with the descent of the scapula which commences during the second month, than that any abnormality in the growth of the scapula should interfere with the ossification of the spine. During the eighth week of intra-uterine life the ribs are already marked out by cartilage, and it cannot be denied that in my case the presence of a cervical rib or of rib-fusion might interfere with the
position of the scapula though not in any way influencing the vertebral development.

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