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DORSAL HEMIVERTEBRA

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A dorsal hemivertebra is taken to mean a vertebra in which the anterior half of the ossification centre in the body has failed to develop.

Another vertebral malformation, a cleft vertebra, has so many features in common with the dorsal hemivertebra, when situated around the thoraco-lumbar junction that occasionally it is called a dorsal hemivertebra, although this is not correct.

Apart from the bipartition, the body of the cleft vertebra has originally been similar to that of the other vertebrae, but, presumably because of the distribution of pressure at this site in the spine, the cleft vertebra is gradually pushed backward and takes on a wedge shape with the pointed end forward.

In the sagittal plane an X-ray film will show identical appearances in the two types of vertebral defect. A more or less wedge-shaped body with subluxation backward in relation to the adjacent vertebrae. This leads to an increased kyphosis or gibbus. The anterior edges of the adjacent vertebrae approach one another, may show compensatory growth, and may at last be as close to each other as two normal vertebrae (*Güntz* 1957, *Junghanns* 1937). Incidentally, it is a characteristic feature that the intervertebral discs adjoining the dorsal hemivertebrae are normal.

Anteroposterior X-ray films will settle the doubt as to which vertebral malformation is concerned, especially when supplemented by tomography. In such a film the cleft vertebra will present itself as two symmetrical triangles on each side of the midline, the smallest angles facing, and separated by a gap of varying size. As a whole, the cleft vertebra is increased in width as compared with the neighbouring vertebrae (*Müller* 1931).

Cleft vertebrae may be situated at any level in the spine and often

co-exist with other vertebral malformations, block vertebrae etc. However, the most common sites are those mentioned above, the upper lumbar and particularly the lower thoracic spine.

The true dorsal hemivertebrae are most often localized in the first and second lumbar vertebra, but may occur in the 3rd lumbar and the 11th or 12th thoracic vertebra. It may be present as the only deformity (*Lindemann* 1931, *Bakke* 1935), and as a link in more widespread diseases, such as achondroplasia (*Caffey* 1957), cretinism and chondrodystrophy (*Schinz, Baensch, Friedl & Uehlinger* 1952, *Bracher* 1933, *Bauer* 1933), *e.g.* in one-third of the cases with the Morquio type and even more often in gargoylism (*Fairbank* 1951). The dorsal hemivertebrae in the two types of chondrodystrophy exhibit characteristic variants of the wedge shape: a pointed "bill" in the middle of the anterior aspect of the vertebra in Morquio's type; in gargoylism the bill is blunt and situated low on the anterior aspect of the vertebra.

There is no doubt that dorsal hemivertebrae, especially solitary ones, are rare, but it is impossible to state the exact incidence. It is important, however, to be aware of their existence and characteristics, especially in order to avoid confusing them with spondylitis. This justifies the publication of the following cases.

CASE REPORTS

Case 1. Case rec. K 19432. A male, now 22 years of age. As early as the age of 6 months, his mother had noted that "his back collapsed" when he was to sit. At first, he was treated elsewhere as a case of rickets, later as spondylitis. Admitted here at the age of 14 months. There was a non-fixed, arcuate gibbus at the level of the upper lumbar spine. No direct or indirect tenderness, no neurological abnormality, and no abscess formation, E.S.R. 5 mm., Mantoux negative. X-rays of the spine: *vide infra.* It was noted that the patient showed faint clinical sequelae of rickets. He had a fairly large rectangular fontanelle, but otherwise he was described as normal. Radiography of all long bones failed to show any signs of rickets, but the metaphyses were widened, massive, and condensed. These changes were interpreted as signs of Møller-Barlow's disease. The patient had light treatment, vitamins C and D and calcium. A few months later the changes in the long bones had distinctly regressed. The spine was treated by a plaster cast for 6 months. After that time, the gibbus was completely unchanged, and so were the X-ray appearances (cf. Figs. 1 and 2): The anteroposterior measurement of the body of the 2nd lumbar vertebra (v. l. II) was 15 mm., while v. l. I measured 20 mm. Its height was the same as that of v. l. I. It was slightly wedged with a rounded, forward directed point and slightly backward displaced, especially in relation to v. l. III. The bone structure was normal, the body smooth with sharp contours, and the intervertebral spaces normal. On the anteroposterior view the vertebra looked completely normal. The patient was discharged two months later.

Fig. 1.

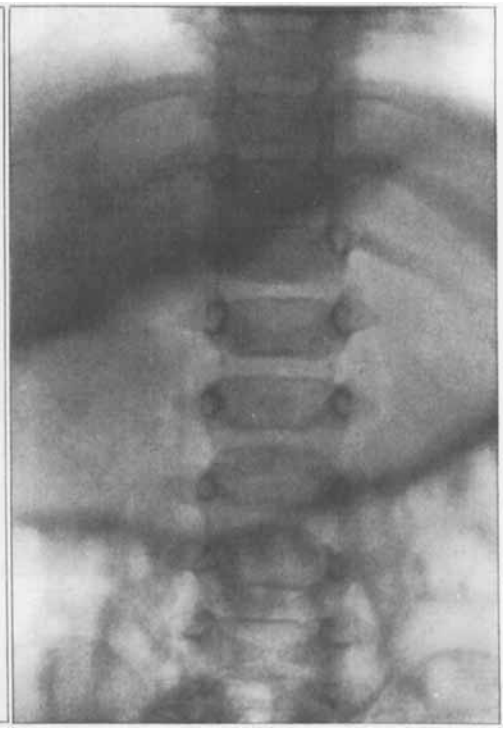

Fig. 2.

At the age of 18 the patient applied to another hospital. He was now a farm hand and complained of backache of many years' duration. Again, the arcuate kyphosis on a level with v. l. I, II, and III was noted. Now, it was somewhat fixed. No other clinical abnormalities. X-ray examination (Figs. 3 and 4) showed that the kyphosis, which had been about 28° at the age of 18 months, had now increased to about 52°. v. l. II had become somewhat more wedged, the backward dislocation was unchanged. The height of the body was equal to that of v. l. III, its anteroposterior measurement 35 mm. as compared with v. l. III which was 50 mm. The intervertebral spaces were normal. There was no compensatory growth of v. l. I or v. l. III, rather a flattening of the lower and upper anterior edges. An unmistakable sign of the increased kyphosis was the finding that the minimum distance between v. l. I and v. l. III had not changed from the age of 18 months to the age of 18 years. The anteroposterior view showed that there was not a qeustion of a cleft vertebra.

Apart from re-training for another occupation, he was not given any treatment.

Case 2. Case rec. K 37750. A boy, aged 12 years. A strong family history of tuberculosis. The patient had "pneumonia" at the age of one year, and thereafter the Mantoux test was positive. His mother noted a gibbus 7 months prior to admission and definitely claimed that it had not been present before that time. She had also

Fig. 3.

Fig. 4.

noted that the patient had got into the habit of immobilizing his spine. He complained of fatigue, but not of pain. No neurological abnormalities.

The patient was admitted to another hospital where his disease was interpreted as tuberculous spondylitis of v. th. X. He was treated by chemotherapy and bed rest, later with a plaster corset, and was transferred to us because of better facilities for education. We found a non-fixed gibbus (Figs. 5 and 6) in the lower thoracic spine. Radiography, supplemented by tomography in two planes (by a Polytome at the Frederiksberg Hospital, Figs. 7 and 8), showed a kyphosis of approx. 50° on a level with v. th. X. At the same site there was a left-convex scoliosis of about 10°. Laterals showed the body of v. th. X to be small and wedged, with the pointed end forward. Its anteroposterior measurement was about 22 mm. as compared with the neighbouring vertebrae 35 mm., while its height was the same as that of the neighbouring vertebrae. There was a slight backward dislocation, most evident in relation to v. th. IX, and the pedicle seemed somewhat wider than on the adjacent vertebrae. The bony structure was normal and the contours sharp and clear. The facing anterior edges of v. th. IX and XI were somewhat flattened, and the distance between them no greater than the distance between the anterior edges of v. th. XI and XII. The intervertebral spaces adjoining the deformed vertebra appeared to be slightly narrowed. However, this was not apparent from the anteroposterior view which showed that v. th. X was a cleft vertebra of increased width and of "butterfly" shape. The right half of the body was smaller than the left, in accordance with the scoliosis. There was distinct compensatory growth of v. th. IX (less distinct of v. th. XI), especially around the centre of the vertebra corresponding to the cleft in v. th. X. The patient was treated by training the spinal muscles.

Fig. 5.

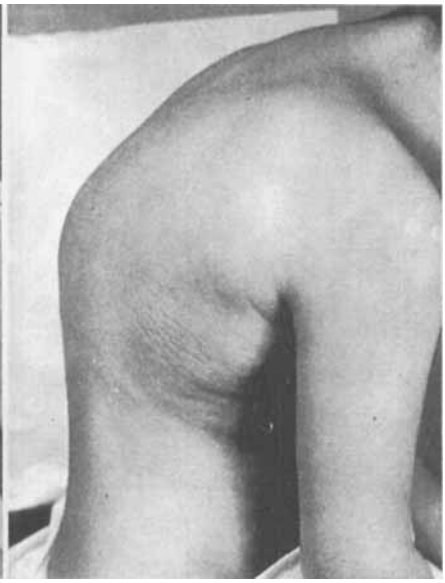

Fig. 6.

Fig. 7.

Fig. 8.

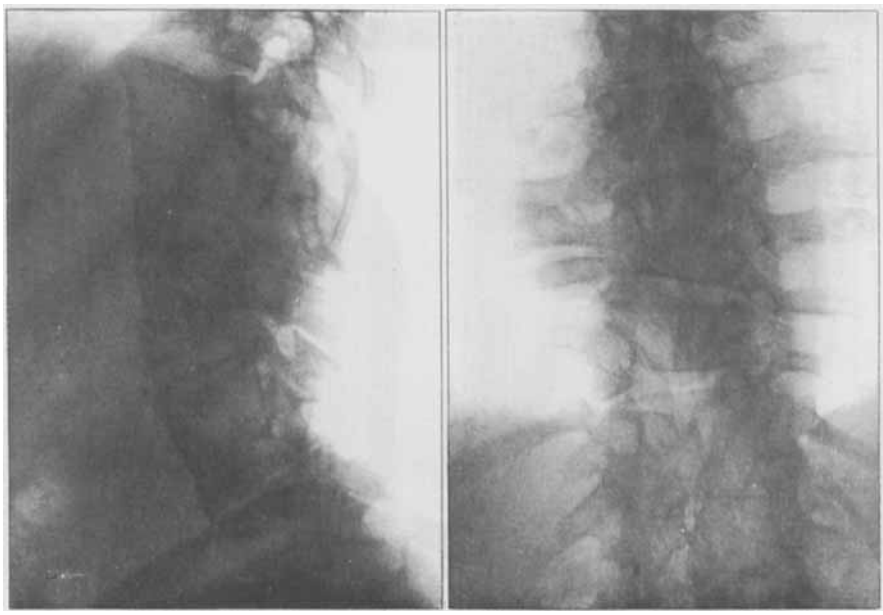

Fig. 9. *Fig. 10.*

Case 3. Case rec. 183042. A female, aged 39, who presented herself because of another disease. She was found to have abundant hairing on the back on a level with the lumbar spine. She had not had backache and there was no abnormal kyphosis.

X-ray examination (Figs. 9 and 10) revealed numerous vertebral deformities, extending from v. th. V to v. l. V. The v. th. X was wedged, greatly backward displaced, so that its anterior edge was 10 mm. behind those of the neighbouring vertebrae, although the measurement from before backward did not differ definitely from the neighbouring vertebrae. Incidentally, this also applied to the height. It was impossible to asses the intervertebral spaces. Marked compensatory growth of v. th. XI. Anteroposterior view showed a cleft vertebra slightly increased in width.

It was noted that other split vertebrae were present, int. al. v. l. IV (not visible on the anteroposterior view). On the lateral view the v. l. IV is wedged, it is true, but not posteriorly displaced and definitely not of an appearance suggesting a dorsal hemivertebra. If anything, it would seem to come within the term wedged vertebra which had been used by several authors.

DISCUSSION

While in Case 1 the diagnosis is a true, congenital dorsal hemivertebra, Cases 2 and 3 are examples of the "false" type of dorsal hemivertebrae, caused by the pressure upon cleft vertebrae.

Case 3 did not give rise to diagnostic difficulties, both owing to the

absence of acute symptoms and because of the numerous other vertebral malformations which immediately pointed into the right direction.

In Case 2, on the other hand, the allegedly acute onset of a gibbus and the possibility of infection led to a diagnosis of tuberculous spondylitis. Not until tomography was done in anteroposterior cuts did the true nature of the disease reveal itself, as this investigation clearly visualized the cleft vertebra and the marked compensatory growth of the adjacent vertebrae. The explanation why the dorsal hemivertebra was not detected until the age of 12 years may be that the kyphosis has developed so gradually that nothing unusual was discovered until the boy started having other symptoms. A sudden "collapse" appears unlikely, as the compensatory growth of the neighbouring vertebrae must have been going on for a long time.

In Case 1 there was also a suspicion of tuberculous spondylitis, but owing to the normal structure and sharp outlines of the vertebra, and the normal intervertebral space, this diagnosis could not be confirmed. Another distinctive factor was the backward subluxation which is an important diagnostic criterion. The appearance of the other vertebrae and the entire clinical picture showed that the dorsal hemivertebra was not a link in a more extensive disease, *e.g.* chondrodystrophy.

It was only in Case 1 that the development could be followed from the time (4 months of age) before the deformed vertebra had been subjected to weight-bearing until a time when the spine had been exposed to great strain. It is remarkable that the dorsal hemivertebra developed and grew to exactly the same extent as the adjacent vertebrae. Only, a slight increase in the wedging was noted simultaneously with a marked increase in the kyphosis.

If the other two patients had been examined at the age of 4 months, it must be assumed that their cleft vertebrae, viewed from the lateral aspect, would have presented an appearance almost identical with that of the other vertebrae.

The kyphosis in Case 2—the 12-year-old boy whose back can hardly have been subjected to any major strain—was equally severe as in Case 1, the 22-year-old farm hand. This indicates that it is the movements of the spine which lead to increased kyphosis rather than actual weight-bearing due to heavy work. The explanation that Case 3 had not developed kyphosis must be the simultaneous development of block vertebrae.

The aetiology of the vertebral malformations is unknown. As a rule,

a "defect" in the notochord is held responsible. The defect which leads to the formation of cleft vertebrae is developmentally earlier than that which causes a dorsal hemivertebra, as the fusion of the originally two hemilateral primordia of the vertebral body occurs before the ossification centres start their development. *Junghanns* assumed that the dorsal hemivertebra arose owing to the absence of the primordium of the anterior one of the two central ossification centres. However, if it is assumed that only one ossification centre is present (*Schinz et al.*), the mechanism of the defect would sooner be failure of vascular ingrowth from the anteerior aspect.

Familiar occurrence of dorsal hemivertebrae has been reported, especially in chondrodystrophy (*Bracher*), and only once in the absence of other skeletal deformities (*Bauer*).

The importance of a really detailed X-ray investigation of unusual vertebral malformations is greatly emphasized by the present case histories. It is important to avoid unnecessary, prolonged treatment, and to advise the patients at an early stage regarding future occupation or possibly a change of occupation.

SUMMARY

Two types of dorsal hemivertebra are described, a "true" type due to failing development of the anterior part of the ossification centre of the vertebral body, and another type due to increasing deformation of a cleft vertebra. Both types are localized around the thoraco-lumbar junction and usually lead to marked kyphosis. Three cases are reported, and the differential diagnosis is discussed.

RESUME

Il existe deux types de demi-vertèbres dorsales, un "vrai" dû au manque de développement de la partie antérieure du noyau osseux du corps vertébral et un autre dû à la déformation grandissante d'une vertèbre fendue. Les deux types sont localisés dans la région de transition thoraco-lombaire et mènent en règle générale à une grave cyphose. Trois cas sont examinés et il est discuté du diagnostic différentiel.

ZUSAMMENFASSUNG

Es gibt zwei Typen von dorsalen Halbwirbeln, eine echte, der eine manglende Entwicklung der vordersten Teiles des Wirbelkörper-

knochenkernes zu Grunde liegt und eine andere, die durch zunehmende Deformierung eines Spaltewirbels hervorgerufen wird. Beide Arten lokalisieren sich in der Gegend des thorako-lumbalen Überganges und führen in der Regel zu schwerer Kyphose. Drei Fälle werden dargestellt und die Differenzialdiagnose wird besprochen.

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