NILS LINDSTRÖM, LUND:

VERTEBRA PLANA CALVÉ

In January 1925 *Calvé* published two cases of a new disease for which he suggested the name of »osteo-chondrite vertébrale infantile«. One of these cases he had seen himself and of another he had obtained particulars from Dr. *Brackett* of Boston. Both patients were boys, 2½ and 7 years old respectively, who came for treatment for symptoms of spondylitis with a gibbus formation in the thoracic region. During the whole period of treatment they were regarded as tuberculous spondylitis and it was only the study of the radiograms that later on led *Calvé* to revise his opinion.

The following are the points on which *Calvé* considered the disease radiologically differed from tuberculous spondylitis:

1) One vertebra only is engaged in the process, and uniformly flattened into to a thin plate. In tbc-spondylitis, on the other hand, the deformity is always formed by at least two vertebrae, destroyed in a wedge-like manner with the intervening intervertebral disc more or less also the seat of destruction.

2) In these cases on the other hand the intervertebral discs are entirely intact.

3) There is even an increased thickness of the cartilage causing *Calvé* to speak of a »néo-formation cartilagineuse«.

4) The vertebral rest yields a denser roentgen shadow indicating a heightened density of bone.

5) Finally regeneration of the vertebra takes place which Calvé considered out of the question in the case of tbc-spondylitis, »l'os tuberculeux ne se régénère pas«. (*Menard* quot. after *Calvé*.)

Calvé emphasized the great similarities existing between this new disease and Perthes' as well as Köhler's diseases.

He held that the bone nucleus in the vertebral body, at this age period surrounded by a solid cartilagenous shell, was affected by a slight inflammation, »serrait atteint de phénomènes inflammatoires à l'étouffée«, whereby it became entirely or partially destroyed. In this period of disease the patient would get pains in the back and a gibbus would form as a result of the cartilagenous shell being compressed after destruction of the contents. Later on the bone nucleus would gradually regenerate and the back regain its normal shape.

In 1927 *Harrenstein* of Amsterdam published two cases of this disease, one of them a boy aged 5 and the other one a girl of the same age, both with symptoms of spondylitis and gibbus formation in the lower thoracic region and with radiograms characteristic of the disease described by *Calvé*. *Harrenstein* was fortunate, however, in securing radiograms in one of these cases during the developmental stage of the disease. These then revealed atrophy of the affected vertebra which was lower than the adjacent ones and had ill-defined outlines. In the centre it showed a zone of denser more shadow-producing tissue. The adjacent intervertebral discs were lower than normal. Four months later the contours of the vertebra seemed to have disappeared so that only the middle calcium-dense portion appeared on the skiagram, at the same time as the adjacent intervertebral spaces had become higher than normal. *Harrenstein* then concluded that the increased intervertebral spaces visible on the radiograms consisted of discs in addition to a much decalcified bone no longer capable of producing any shadow. For this view he also found support in the subsequent regeneration of the vertebra brought about by deposition of lime in the previously much decalcified bone. In other respects *Harrenstein* shared *Calvé*'s opinion that it was a question here of an independent earlier unobserved disease which he compared with Perthes' and Köhler's diseases. On account of the appearance of the vertebra *Harrenstein* suggested it should be named »vertebra plana« which must also be considered an adequate term. Since however

a number of other conditions exist with flattened vertebrae the name of »vertebra plana Calvé« has later been generally adopted.

At the Lund Orthopaedic Clinic we have for a long time been having the opportunity of following a case of vertebra plana Calvé and since it in several respects is of great interest I propose to place in on record.

It is the case of a boy, born June 11 1923. He was admitted to the Lund Orthopaedic Clinic on June 18 1927, thus at the age of 4. There

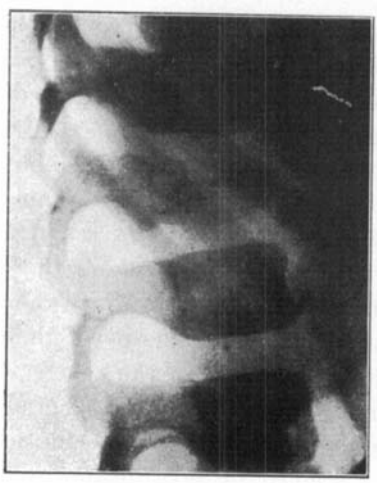

Fig. 1.

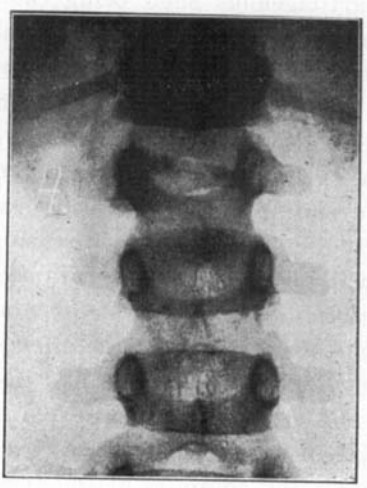

Fig. 2.

was no known exposure to tuberculosis and while under treatment for bronchitis in another hospital for a week in 1927 he twice showed a negative Pirquet. For a fortnight before admission he had complained of pains in the back and for the past seven days his parents had noticed he held his back stiffly and that he was somewhat crooked.

Condition on adm. June 18 1927.—General condition good. Wellbuilt boy, well-nourished. Internal organs, nil abnormal.

Carries his back very stiffly. When he is to pick up something from the floor he squats. Mild kyphosis and a slight scoliosis of the lumbar spine. No definite tenderness on percussion of spinous processes.

Temp. normal. Pirquet neg. Mantoux 0.01 mgm. positive.

A note on Oct. 27 states that the knee-jerks were brisk but that the plantar reflex was flexor.

Rtg. ex. of June 21 1927 shows flattening of L. II, particularly of its

front part, giving to the vertebra on a lateral picture the appearance of a figure tapering in front. Well-marked decalcification with a denser zone in the centre, a few millimetres broad. Adjacent intervertebral discs are somewhat reduced in height. On an anteroposterior picture the left half of the vertebra appears much lower. In this projection, too, there is a denser zone in the centre of the vertebra surrounded on both sides by a narrow portion of decalcified bone. (See fig. 1 and 2).

Fig. 3.

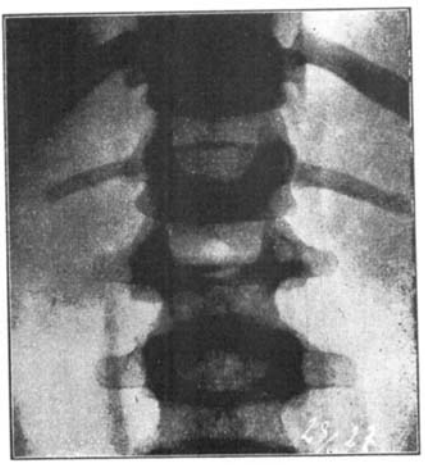

Fig. 4.

Rtg. ex. of Oct. 28 1927 shows much more increased flattening of L. II. There only remains now a narrow dense portion of the vertebra which in a lateral view projects in front of adjacent vertebrae. Neighbouring intervertebral spaces are higher than normal. (See fig. 3 and 4).

Rtg. Jan. 11 1928 shows a slight yet obvious regeneration of the vertebra. (See fig. 5 and 6). This regeneration appears still more clearly on July 2 1928 and on Dec. 27 1928 the vertebra seems to measure about its normal height. In August 1929 the vertebra shows normal bone structure. Regeneration is proceeding year by year so that on Aug. 17 1931 the vertebra is reformed to about $^2/_3$ of its size and has good bone-structure. In a lateral view it is still seen to project somewhat beyond neighbouring vertebrae and in the antero-posterior view its left part is still slightly lower though the difference is almost equalized. The intervertebral spaces still seem rather high. (See fig. 7 and 8).

Fig. 5.

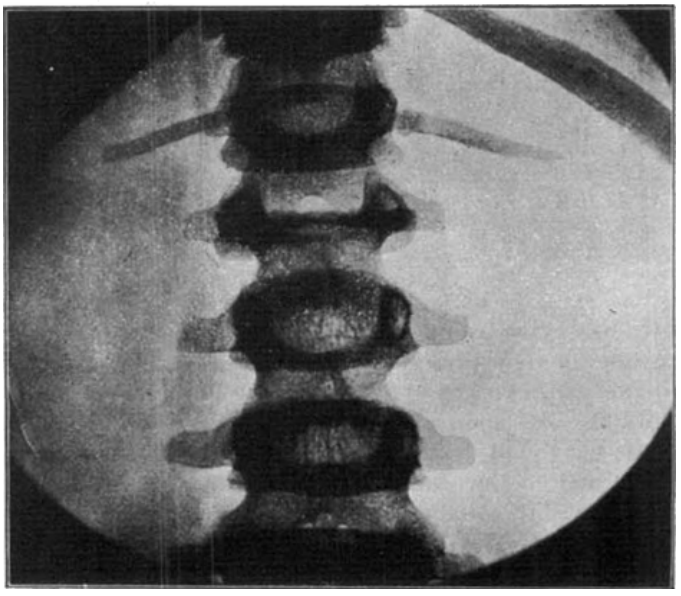

Fig. 6.

The patient was first treated in plaster bed for 7 months until Jan. 25 1928. He was then given a plaster jacket till July 11 1930 when he was fitted with a leather jacket which he wore till August 1931.

In January 1928 it is stated in the case record that the kyphosis is nearly gone and that there is good mobility in the back. While he was wearing a jacket he had no symptoms whatever and in August 1931 there

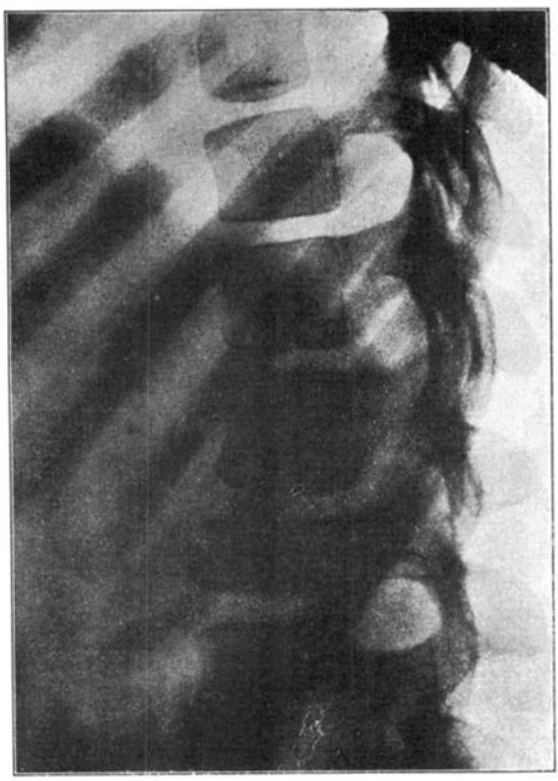

Fig. 7.

is a note saying that he had free mobility in his back but still an indication of kyphosis at the old place.

On April 4 of this year I had the patient in hospital for after-examination. Since leaving off wearing the jacket in 1931 he has not had the slightest trouble with his back. He has moved about freely and taken part in the gymnastic class in school without any difficulty.

No deformity of the back at present. No tenderness on percussion over vertebrae. Free mobility of back without pain. Reflexes normal.

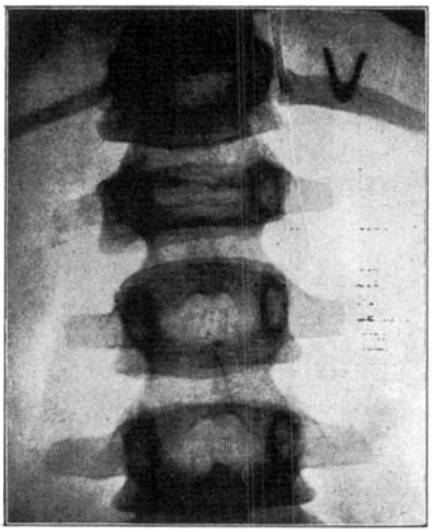

Fig. 8.

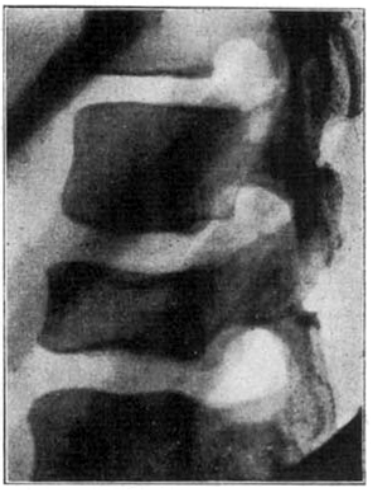

Fig. 9.

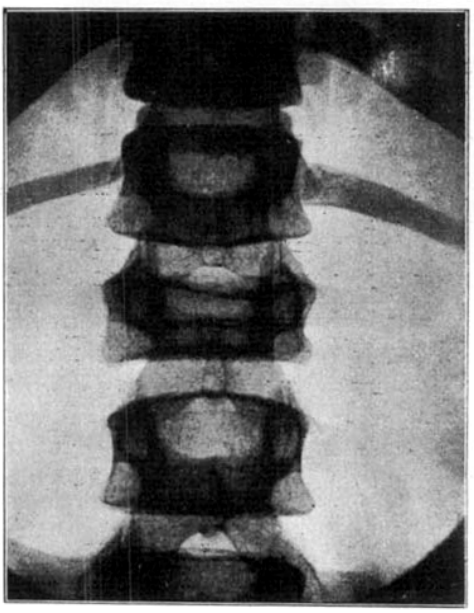

Fig. 10.

Rtg. of April 4 shows still further regeneration of the vertebra which now has about ¾ of its normal height. Calcium contents and bone structure normal. In a lateral view the vertebra has a bi-concave shape and is still projecting somewhat beyond neighbouring vertebrae. In an antero-posterior view the vertebra is now very nearly symmetrical in shape. The intervertebral spaces are still, if possible, somewhat broader than normal. (See fig. 9 and 10).

The case is of interest because radiologically one had the opportunity to follow the disease during its developmental stage and subsequently during the period of regeneration that has gradually proceeded in the course of the past 7 years. In the past only *Harrenstein, Hanson* and *Lamy* have been able to follow the disease during its developmental period. Thus *Hanson* has a normal radiogram and one 40 days later in which the vertebra can be seen collapsed into a plate a few millimetres in height, while *Harrenstein* and *Lamy,* like ourselves, have radiograms from the developmental period. In no previous case has one followed the regeneration roentgenologically during such a long period as we have had the opportunity of doing. In our case this regeneration began after a couple of months and still seems to be in progress after 7 years, the height of the vertebra has, at any rate, increased since 1931.

In the course of years a number of cases have been published and from the world's literature I have managed to collect 24 looked upon as vertebra plana Calvé. Some of these, however, must be excluded as certainly not belonging to this category.

So do we find *Buchman* in the January number of The Journal of Bone and Joint Surgery 1927 describing two cases which he considers belonging to vertebra plana Calvé. In one of these cases roentgen shows changes in the upper thoracic region with, in the lateral picture, several vertebrae compressed and wedge-shaped. These vertebrae have regular outlines with an increased density of the upper and lower limiting borders, and the intervertebral spaces are described as somewhat increased in relation to the flattened vertebrae. I am unable to find that the changes shown in these skiagrams justify the case being referred to the group of vertebra plana Calvé. Nor in the second case would there seem to be any roentgen appearances characteristic of

vertebra plana Calvé. It is the case of a girl who as early as at the age of 20 months was treated for a scoliosis which was regarded as congenital. At the age of 11 she was admitted to hospital complaining of her back. She had then an S-shaped scoliosis with changes above all of D. XII and L.I which had irregular outlines and on antero-posterior pictures proved to be wedge-shaped. L.V. was asymmetric in shape and all the dorsal vertebrae showed irregular outlines with compressed intervertebral discs. To my mind this case seems most like a scoliosis as a result of some developmental disturbance.

In October 1927 *Boorstein* likewise published in The Journal of Bone and Joint Surgery two cases which according to his opinion are to be referred to as vertebra plana Calvé. The first case is however a youth, aged 17, with a typical kyphosis dorsalis juvenilis (*Scheuermann*). The second case is that of a girl, aged 2, with mild symptoms of spondylitis. The first radiogram shows a slight diminution of L.III with intervertebral discs intact. A picture 15 months later shows progression of the lesion so that L.III is nearly entirely destroyed and L.II and L.IV show slight erosions. After another two years L.III is entirely destroyed and a large defect is seen in the lower half of L.II causing this vertebra to be deformed in a wedge-like manner. Neither clinically nor roentgenologically are there any symptoms here that can justify a diagnosis of vertebra plana Calvé, the case instead being undoubtedly a mild form of tuberculous spondylitis.

It seems strange that a great many authors would have included these cases without reservation. It is really only *Federschmidt* and above all *Hanson* who have made any objection, the latter in his publication in Acta Chirurgica Scandinavica 1930.

Among the others I have been unable to check a case of *Gallie* who, according to a statement by *Buchman,* he described before the congress of the American Orthopaedic Association in May 1925. In an article appearing in La Presse Thermale et Climatique 1931 *Calvé* refers to an unpublished case of *Platt* of Manchester besides a case of *Kleinberg.* The latter case, how-

ever, would seem to be the same as Buchman's second case which during an earlier phase of the disease had been examined by *Kleinberg*. In the discussion that followed on *Panner*'s demonstration of his case *Heyerdahl* in Norway said he had observed a similar case in a 8-year old boy but this had not been published either.

Out of the 24 cases published in the literature I thus find that after having excluded some which are certain not to belong to the category concerned and further sifted those unpublished and therefore uncontrollable, there remain but 16 cases. If adding our own case there would thus exist so far 17 cases where vertebra plana Calvé could be justified as a diagnosis, even if the disease in one or two of them is not fully typical. *Federschmidt* maintains, however, that the disease occurs much more commonly than indicated by these figures and that a great many cases remain undiagnosed on account of lack of knowledge of the disease. Since however the disease, roentgenologically, is so distinct from tuberculous spondylitis, which diagnosis would practically cover the mistaken cases, it seems to me unlikely that these cases would include any greater number of vertebra plana Calvé. The disease in question therefore must be considered very rare.

Out of these 17 cases there are 11 boys and 6 girls, thus here as in Perthes' disease a definite preponderance among boys although of course the cases observed are as yet too few to allow any definite conclusions being drawn. In 11 of the cases the disease began between 4 and 7 years of age. In 4 cases the children were above this age viz. in a case of *Löhr* where the disease began at 8 years of age, in one of Schrader at 12 years, one of *Panner* at 14 and in one of *Janzen* at 15 years of age. With regard to the two latter cases the age here may naturally make us somewhat doubtful about the diagnosis, still more so as the course was otherwise not typical either. In *Janzen*'s case there thus arose nerve symptoms in the form of exaggerated reflexes and a positive Babinsky, and in *Panner*'s case there was after 8 years no regeneration of the affected vertebra which is otherwise regarded as the most characteristic feature of the

disease. Since however the skiagrams in both cases may otherwise be regarded in favour of vertebra plana Calvé I have included them, particularly since on account of the scarcity of cases hitherto observed we are unable as yet to know the different variations under which the disease may appear. In one of Calvé's cases the disease commenced at 2½ years of age and in one of Federschmidt's at the age of 3. Even if the disease may commence between the ages of 2 and 15 years, the age of predilection, like in Perthes' and Köhler's diseases, would seem to lie between 4 and 7 years.

In 15 cases one vertebra only was affected, the lesions being located in the lumbar or lower thoracic spine. In one case 2 vertebrae were affected and in one case, finally, several vertebrae were gradually affected.

Pirquet has been negative in 9 and positive in 3 cases. In the rest the tuberculin reaction came out negative in 3 cases and in one case there is no statement of any tuberculin test. Mantoux was carried out in 5 cases, of which 4 proved negative and one positive. Wassermann proved negative in all the 11 cases tested. The temperature has generally been normal; in 4 cases however it was stated as subfebrile.

In all the cases the patients have had symptoms which clinically accord well with tuberculous spondylitis. That there can be no question of tuberculosis is shown however by the great number of negative tuberculin reactions but clinically it is thus impossible to distinguish in the individual case vertebra plana Calvé from tuberculous spondylitis.

In one case definite nerve symptoms were present in the form of exaggerated reflexes and a positive Babinsky but as a general rule no nerve symptoms seem to occur.

As is evident from *Harrenstein's* case and our own, roentgenologically, there will be found in the earlier stages of the disease an initial flattening of the vertebra which is markedly decalcified, shows ill-defined boundaries and centrally presents a denser zone a few millimetres wide. In the course of further development the vertebra is getting rapidly flattened out so that only the middle calciumdense portion remains. Concurrent-

ly with this we get a gradually increasing breadth of the intervertebral spaces above and below the affected vertebra which fits in well with Harrenstein's view that the increased intervertebral spaces are formed by the intervertebral disc plus decalcified bone. A characteristic feature of vertebra plana is the regeneration setting in later and which in our case commenced already after a couple of months and which has subsequently been going on for 7 years. Simultaneously with the regeneration the intervertebral spaces resume very nearly their normal height which of course supports *Harrenstein's* view that regeneration would occur as a result of lime being deposited in the decalcified bone, earlier invisible under roentgen. This theory however seems insufficient since in our case we found a considerable regeneration even after the intervertebral spaces had become almost normal. Like in some other cases we have observed also in our case how the flattened vertebra on a lateral picture projects somewhat beyond the adjacent vertebrae; this has been interpreted as a sign of compression because of the diminished resistance of the decalcified vertebra. Roentgenologically we may thus find characteristic appearances of vertebra plana Calvé but similar pictures may at times be seen in tuberculous spondylitis in children with discs relatively intact and with an almost flat vertebra; it will thus be the course of regeneration of the vertebra that will definitely settle the matter. It is true that regeneration may sometimes occur in tuberculosis but this will never reach the extent as in vertebra plana Calvé.

As to the prognosis this must be considered satisfactory since evidently symptoms generally disappear after a short time. In one case only have persistent kyphosis and pains in the back been recorded.

As long there are no autopsical findings on record of vertebra plana we are unable to state definitely what pathological changes are at the back of the disease. However, as I have already stated, it bears great similarities to malacia of various localities, above all then Köhler's disease of the scaphoid of the foot. *Panner* is nevertheless pointing out certain dissimilarities between this disease and vertebra plana. So is in Köhler's disease the

distance between the astragalus and the cuneiform always normal and complete recovery gradually takes place, a thing never as yet observed in vertebra plana. This may be explained however as due to the greater loading to which the vertebra is subjected, more so since the process is generally localized to the lumbar or lower dorsal spine which would handicap the healing process in vertebra plana. As is pointed out by Federschmidt this does not bring us any nearer to the causal genesis, this being still obscure so far the local malaciae are concerned.

In all the cases treatment seems to have been fixation in one way or the other, generally by plaster bed or jacket. In the cases first published who during their whole period of disease were regarded as tuberculous spondylitis the fixation was a long one; subsequently however the time was shortened so that the patients became free with abatement of symptoms. This seems to me however a wrong procedure because if a failing regeneration is due to the loading, then the period of fixation should be prolonged as much as possible. Up till now after-examinations carried out have been too incomplete to enable us to judge from the result to what extent the regeneration is influenced by the period of fixation but this much can be said that in the two cases, Hanson's and our own, where the most powerful regeneration was observed early and prolonged fixation had been resorted to.

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