DIMINISHED SEGMENTATION OR PREMATURE OSSIFICATION OF THE STERNUM IN CONGENITAL HEART DISEASE

BY

L. ANDRÉN AND P. HALL

From the Roentgen Department and the Medical Department, Malmö General Hospital, Malmö, Sweden

Received April 25, 1960

Bony union of the segments of the stenum starts in early childhood but is normally not complete before the 15th year of life (Ashley, 1954). We have observed early fusion or defective segmentation of the sternum in six patients with congenital heart disease and in one with a provisional diagnosis of heart disease. Subsequent search of the roentgen files of 1100 patients admitted because of suspected chest disease failed to reveal any cases of diminished segmentation but, in addition to the above-mentioned seven cases, we have seen a number of patients with less striking defective segmentation of the sternum, often in association with congenital heart disease. This group is being studied and will be the subject of a future publication. Diminished segmentation of the sternum has also been noticed in another 7 cases without detectable cardiovascular disease.

In 1956, Monnet *et al.* described a case in which autopsy revealed partial transposition and an aberrant pulmonary vein as well as a sternal anomaly—*ossification prématurée du sternum*. Currarino *et al.* (1958) reported 3 examples of this condition; one patient had a large septal defect with a wide ductus arteriosus confirmed by autopsy, another had radiological evidence of an aberrant right subclavian artery, and the third had no heart disease. These authors also published seven other cases with less severe sternal anomalies, three of them with vascular deformities.

Case Reports

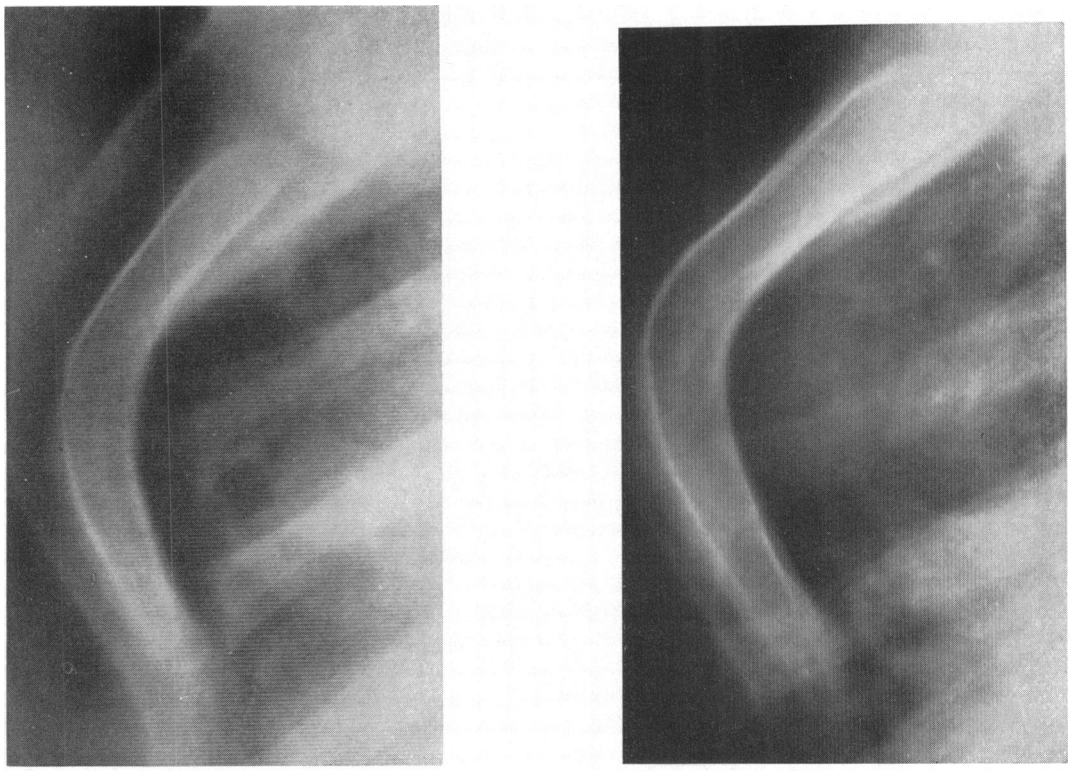
Case 1. A girl, aged 15, was shown by clinical examination and cardiac catheterization to have atrial and ventricular septal defects with pulmonary hypertension. Examination of X-ray films taken at the age of 9 months showed that the manubrium and the sternum were almost completely fused and that the manubrium showed no evidence of the normal segmentation. The forward curvature of the sternum was somewhat more pronounced, and at the age of 5 years it was severe (Fig. 1).

Case 2. A boy with Fallot's tetralogy had a Blalock operation at the age of 5 years and a further operation, when aged 14, under extracorporeal circulation, from which he died. Post-mortem examination confirmed the diagnosis of Fallot's tetralogy. Examination of X-ray films taken at 5 years showed that the manubrium and body of the sternum were fused and that segmentation of the latter was hardly discernible. The sternum was straight (Fig. 2 a).

Case 3. A boy, aged 6, was shown by autopsy to have an atrial septal defect and slight pulmonary stenosis. Examination of X-ray films taken at the age of 4 years showed fusion of the manubrium and body of the sternum with a slight forward angulation (Fig. 2 b).

Case 4. A boy, aged 10, was admitted to hospital for correction of syndactylia of the left hand. Roentgen examination showed a normal-sized heart displaced to the right and electrocardiography showed signs of right-sided hypertrophy. Physical examination showed a systolic murmur (grade 2) over the apex. A diagnosis was not made. Examination of X-ray films taken at 6 years of age showed a short, fused sternum with a midline forward bulge.

Case 5. A man, aged 36, had slight pulmonary stenosis confirmed by catheterization and angiocardiography. Examination X-ray films showed complete fusion and marked forward angulation of the sternum.



a b

FIG. 1.—X-ray of sternum in Case 1, showing fusion of the manubrium and body of the sternum with forward curvature (a) at 2 years of age and (b) at 5 years.

Case 6. Autopsy of a boy, aged 3 weeks, showed aortic valve atresia, a large ductus ateriosus, patent foramen ovale, right ventricular hypertrophy, and left ventricular hypoplasia. Both thumbs showed supernumerary phalanges. Examination of the X-ray films taken at 18 days showed fusion of the three nuclei of the manubrium and slight forward angulation of the sternum.

Case 7. A girl, aged 14, was known to have a vascular anomaly of the renal vessels. The blood pressure was 150/100 mm. Hg, and the electrocardiogram confirmed left ventricular hypertrophy. Rœntgen examination showed slight forward angulation of the sternum.

DISCUSSION

Maldevelopment of the sternum appears to be one of the commonest malformations associated with congenital heart disease. It is surprising that it is not more widely known in view of the importance of pre-operative diagnosis in congenital heart disease, particularly during the last 15 years. Various explanations may be offered for the anomaly having passed unobserved: (a) it has been variously called funnel chest or "voussure", that is a bulging of the precordium due to hypertrophy and dilatation of the heart during childhood; (b) it has been regarded as a sequela of rickets; (c) it has not been observed roentgenologically because cardiac examinations do not always include lateral views; and (d) the sternum is seldom the seat of disease and is therefore not often examined.

Three of our cases had been diagnosed as "voussure," and one as funnel chest for which he was operated upon in 1951. However, re-examination of the X-rays files of 80 other patients with funnel chest revealed no further sternal anomalies of this type.

Descriptions and illustrations of sternal anomalies attributed to rickets have been published but

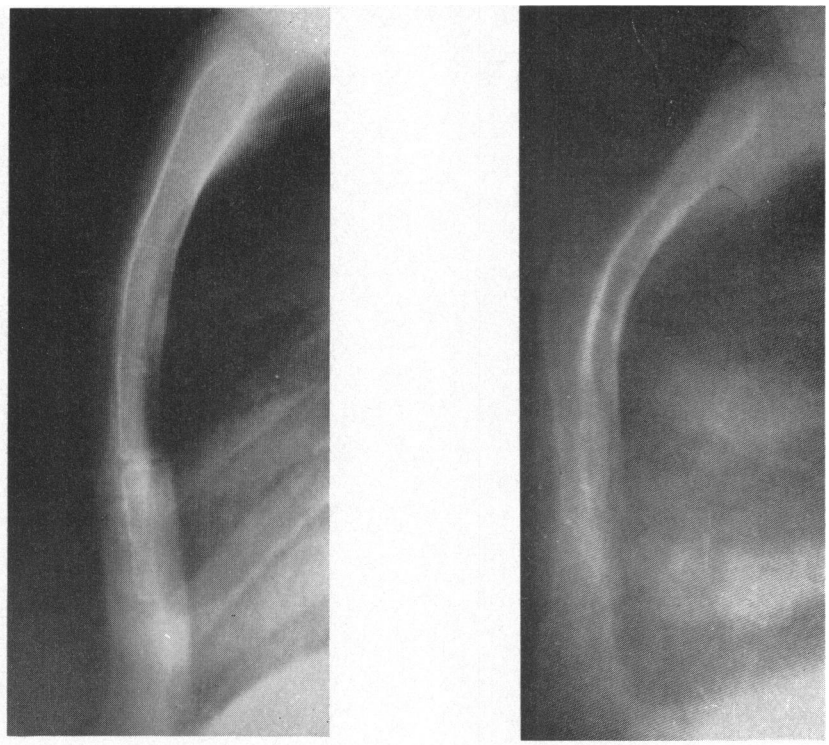


FIG. 2.—X-ray of sternum, showing fusion of the manubrium and body of the sternum; (a) in Case 2 at 5 years of age, and (b) in Case 3 at 4 years of age.

these are probable manifestations of defective segmentation (Zimmer, 1939). Manuals on congenital heart disease contain reproductions of frontal and oblique roentgenograms of the chest but rarely of lateral views, because such views are not generally used in examinations of congenital heart disease. A genetic factor could not be detected in any of our cases. The condition is therefore probably due to some exogenous intrauterine injury. Since the frequency of sternal anomalies appears to vary with the type of congenital heart disease, being more common in the presence of septal defects than with coarctation or pure pulmonary stenosis, investigation of the underlying mechanism is desirable.

SUMMARY

Diminished segmentation or premature ossification of the sternum is one of the most common anomalies seen in association with congenital heart lesions. The malformation is readily recognized in lateral chest X-rays. The finding of such an anomaly indicates examination for congenital heart disease.

REFERENCES

Ashley, G. T. (1954). *Thorax*, **9**, 1959.
Currarino, G., and Silverman, F. (1958). *Radiology*, **70**, 532.
Monnet, P., Gravier, J., Gauthier, J., and Verney, R. (1956). *Pédiatric*, **11**, 95.
Zimmer, E. A. (1939). *Das Brustbein und seine Gelenke Verlag*. Georg Thieme, Leipzig.