ECTOPIA CORDIS CUM STERNI FISSURA

BY

C. E. KELLETT, M.D., M.R.C.P.

The Royal Victoria Infirmary, Newcastle-on-Tyne.

Examples of ectopia cordis are relatively rare, but so striking that they have long been recognized and described, and so have accumulated. In 1818 these cases were classified by Weese¹ into three main groups; ectopia cordis cum sterni fissura, ectopia supra-thoracica and ectopia sub-thoracica, and in summaries this classification has in the main been adhered to. Despite this, cases recorded are as a rule simply termed examples of ectopia cordis, and confusion has been increased by this term in several instances having been so extended as to include cases of dextero-cardia. This paper is concerned with a small group of cases in which, in association with a divided sternum, the heart lies completely outside the body, 'As if,' writes Ignatius de Torres,² 'the heart, not bearing so close a confinement, burst through the breast, and, having broke the sternum, appeared on the outside.' The case to which Torres refers is the one that was most graphically described by Martinez³ in 1723. Subsequent cases have been described by Walter⁴, Tourtelle⁵, Cruveilhier and Monod⁶, Jones⁷, Daniell⁸, Schlesinger⁹, Barnardo¹⁰, Lilwall-Cormac¹¹, Matteucci¹², Ellis¹³, Greiffenberg¹⁴, Rolland¹⁵, Cosgrove and St. George¹⁶, Cutler and Wilens¹⁷, Bloch¹⁸, Siemens¹⁹, Ledényi²⁰.

Case report.

The patient here reported represents the nineteenth in this short series (fig. 1).

R.S. was admitted into the Royal Victoria Infirmary, Newcastle, under the care of Dr. George Hall, on 9.9.35. He was barely a day old, born of healthy parents, the delivery being normal, and pregnancy uneventful. He was the second child and his elder brother was normal, as were also his cousins. He died on the evening of the next day, having been in hospital forty-eight hours. As shown by the photographs, he was well nourished and in excellent general condition apart from a gross hare lip and the pulsating tumour which lay in the midline of his chest. This consisted of the heart which was completely outside the thorax, was foetal in type, and lay vertical to the thoracic wall, the right ventricle facing the chin and the left the toes; at the base of the heart lay the two auricles, the left auricular appendage to the left and the right to right, the vessels of the heart forming the pedicle. With each respiration the heart was pulled into a more vertical position and drawn in, as was clearly demonstrated by a plethysmograph tracing.

As was previously noted by Martinez, handling the heart appeared to distress the child, but when left alone he appeared to be perfectly contented, took nourishment readily from a spoon, and was only very slightly cyanosed. In view of the previous failures reported, no attempt

at radical cure was made, and the child was made as comfortable as possible, the heart being enveloped in gauze soaked with warm saline.

Post-mortem examination ascribed his death to adhesive pericarditis, and Prof. Bramble Green kindly supplied the following anatomical report on the child :—

FIG. 1.

Examination of the intact specimen showed that the heart, which was forming a marked projection in the middle line of the anterior wall of the thorax, was uncovered by fibrous pericardium, and that the skin was directly continuous with the surface of the heart or epicardium. The skin edge at the line of continuity with epicardium or visceral serous pericardium was raised into a prominent collar-like ridge. The specimen was dissected by removal of the soft parts from the surface of the chest wall and cleaning the great vessels entering and leaving the heart,

THE HEART (fig. 2). This consisted of a large ventricular part continuous above with the aorta and pulmonary artery, and small atrial portions projecting on each side of the ventricular part. There was a slight groove on the ventricular part which indicated its separation into right and left cavities. Subsequent examination of the interior

FIG. 2.

confirmed the view that the right ventricle was placed anteriorly and above and the left posteriorly and below. The right atrium was larger than the left atrium, and the left horn of the sinus venosus was persistent.

INTERIOR OF THE HEART. The interior of the right atrium showed a normal atrio-ventricular foramen, and normal pectinate muscles.

A well-marked vertical ridge, starting above and passing down behind the orifice of the superior vena cava, seemed to separate the atrium proper from the sinus venarum or right part of the sinus venosus. There was within the latter cavity the orifice of the inferior vena cava with its valve, a small but patent foramen ovale, and below this a large opening leading into the left horn of the sinus venosus.

The interior of the left atrium presented no unusual features.

The interior of the right ventricle presented a U-shaped cavity, the descending limb passing downwards and to the left from the atrio-ventricular orifice and the ascending limb passing upwards to the orifice of the pulmonary artery. The lower muscular part of the interventricular septum was complete and ended above in a concave upper border. Above this there was a communication with the upper part of the left ventricle where the aortic orifice was plainly seen. Examination of the interior of the left atrium and left ventricle confirmed the deficiencies of the septa noted above.

THE GREAT VESSELS. The aorta appeared to be normal in its arrangement and in the arrangement of the branches which arose from it, innominate, left common carotid and left subclavian.

The veins entering the heart from above were the right and left superior venae cavae. The right vena cava was definitely larger than the left and opened into the posterior part of the right atrium. It received the azygos vein. The left vena cava opened into the left horn of the sinus venosus and also received a vein of the azygos system. The two venae cavae were joined just below the thyroid gland by the transverse communication which normally should form the left innominate vein. Two large inferior thyroid veins joined this vessel.

The pulmonary artery appeared to be normal as to its division, but there was no communication (ductus arteriosus) with the arch of the aorta. The other vessels, as far as could be seen, also seemed to be normal.

The condition of the skeleton of the anterior chest wall was interesting. There were two sternal bars widely separated above, each of which was joined to the first seven costal cartilages and articulated above with the clavicle. The lower ends of these sternal bars were joined by a cross-piece which formed an inferior boundary to the aperture through which the heart protruded.

SUMMARY.

The main anatomical features of the specimen were :—

- (1) Complete extrusion of the heart from the thorax.
- (2) Absence of the parietal pericardium.
- (3) Incomplete development of the ventricular and atrial septa.
- (4) Persistence of the left horn of the sinus venosus.
- (5) Absence of the ductus arteriosus.
- (6) Developmental arrest of sternum.

Discussion.

It is significant that the description by Martinez had almost applied to this child or to the majority of children in this series, for, with the exception of the child reported by Matteucci, whose heart was apparently covered by skin and a normal pericardium, and those by Cosgrove and Ledényi, who had died some time before birth, and in whom there

were other gross abnormalities, these cases all have a great deal in common.

As a rule pregnancy was uneventful, though in several instances^{15, 10, 5, 7}, the child was born prematurely. Moreover, only once¹⁵ did the child die in the course of delivery, owing, it was thought, to pressure on the exposed heart, and this child was born prematurely at six-and-a-half months, and, it so happened, one of twins, the other twin, a dissimilar one, being normal. Boys are more commonly affected than girls, for ten of this series were recorded as being male and only six female, and it seems probable that all races of man are equally affected. This abnormality is not infrequently met with in animals. Though one child recently lived for as long as eight days¹⁸, as a rule such children live for a bare one or two days, the average duration of life being thirty-six hours. Recently there has been an attempt in two cases^{17, 18} to prolong their life by restoring the heart to the chest cavity and covering it over with skin-flaps. In both instances the action of the heart immediately became so embarrassed as a result of external pressure that it was necessary to cut the sutures and release the heart. The most successful treatment at present, then, would still appear to be with a few modifications, that introduced by Martinez, who had made in imitation of the pericardium a little chest of pliant osiers and around this draped a small linen cloth soaked in spirits of wine and melted butter.

Permission for an autopsy in the first-reported case was obtained only after repeated entreaties. In some of the subsequent cases complete examinations were not carried out. With the exception of the case already mentioned, reported by Matteucci, the pericardial sac appears invariably to have been lacking in these children, the parietal pericardium alone covering the heart, and in the majority of instances forming a prominent line of junction with the skin. Frequently^{4, 8, 9, 10, 14} the foramen ovale was patent, and in these cases, with one exception¹⁰, the inter-ventricular septum was also incomplete, while the presence of two superior venae cavae was noticed in several cases^{4, 9, 14, 16} in addition to the one here reported. As a rule the fine general condition of the child has been stressed, but the hare lip and cleft palate, which was so prominent a feature in this child, are also present in some degree or other in several of the series^{4, 11, 12, 16, 19}, and were present to a gross extent in the two babies^{16, 20} previously noted, who were born dead.

Electrocardiograms.

In only one patient, prior to that reported here, have electrocardiograms been taken, and Bloch¹⁸ contents himself with stating that they revealed a normal pulse rate of 130-140 beats per minute. It was felt, therefore, that in this child an attempt should be made to confirm the recent work by Barker, McLeod, and Alexander²¹ on direct stimulation of the heart muscle. As might have been anticipated, since the

heart lay right outside the child's body, electrocardiograms taken from the ordinary leads revealed very little indeed, the heart itself causing scarcely any deflexion at all. Use was therefore made of non-polarizable electrodes and a series of tracings made from direct leads taken from the heart. Such curves have been termed by Samojloff²² electrograms in order to distinguish them from the electrocardiograms taken from indirect leads. In any consideration of these it is important to bear in mind their fundamental differences from the electrocardiogram, for, whereas by virtue of their broad contacts, which are constituted by the tissues surrounding the heart on all sides, the electrocardiographic curves represent much more in their due proportions the activity-effects of all the muscle of the chambers, the electrogram represents simply the result of differences in potential in relatively small areas of muscle lying immediately in contact with the two electrodes. Many of the characteristics of the electrocardiogram appear to depend upon the nature of the contacts rather than upon the heart itself, so that, as shown by Kountz, Prinzmetal and Koenig²³, the dog's heart suitably placed in the human pericardial cavity may yield with the normal leads electrocardiograms 'similar to those obtained with the revived beating human heart.' Though, then, another paper²⁴ of theirs suggests that electrocardiograms taken from the human heart may not in the future be so rare as at first sight one might expect, it would seem reasonable to believe that a great deal of the experimental work done by Craib²⁵, Samojloff²², Woronzow²⁶, Lewis²⁷ and his co-workers in the animal is applicable to the interpretation of electrograms in man, and more particularly to those of this child.

The electrograms in this case were taken as follows:—

As is more clearly shown in fig. 2, a definite sulcus separated the apices of the two ventricles. Three different leads were therefore employed, the first being from the right apex and the left base, the second from the left apex and the right base, and the third from the lateral extremities of the right and left auricles. Non-polarizable electrodes were used. They were of the form described by Gotch³⁰ and employed by Lewis²⁷, with the exception that contact with the surface of the heart was made by means of camel-hair brushes dipped in the kaolin-saline paste and protruding through the end of the glass tube. Electrograms (fig. 3) taken from these leads are here designated a, b and c. The electrograms d and e were taken from the same leads as a and b respectively, but differ in that prior to their taking cold was applied to the tip of the left ventricle in the first electrogram and to the right in the second. This was in both instances effected by means of a test-tube filled with cold water and applied for 30 seconds before the electrogram was made. The sixth (f) tracing represents a normal electrocardiogram (Lead 11) taken from this child, and the seventh (g) an electrocardiogram taken with one lead on the left ventricle of the child and the other on the lower skin of the lower part of the abdomen.

It is apparent that these curves differ from those obtained from the ordinary leads, and their interpretation is rendered peculiarly difficult in that it was impossible in this case to take normal electrocardiograms

A

B

C

D

FIG. 3.

to act as a control. Moreover, the area of the electrodes in contact with the heart is relatively large compared to, for instance, those employed by Lewis who used electrodes having a contact area of 1.5-2 mm. in diameter, whereas the diameter of the ones we employed were from two to three times greater. Furthermore, these electrodes were placed at a considerable distance from one another. Now, as Craib²⁵ has shown, in such circumstances a peculiarly complicated curve may be expected, for such a curve represents not merely the resultant of changes in potential due to processes which have reached the two electrodes at approximately the same time, but rather a resultant due to processes which will have been initiated at different times in the two electrodes.

A further difficulty inherent in all considerations of electrograms lies in the fact that, as Lewis²⁸ points out, curves from electrodes applied directly to cardiac muscle comprise deflexions of two kinds.

‘The chief deflexions are those which result from the arrival of the excitation process immediately beneath the contacts; these are termed intrinsic. They are deflexions, which represent relatively large electrical potentials, and they have correspondingly large amplitudes. (Exceptionally the intrinsic deflection is not the most prominent in the electrogram.) The deflexions of the second order are those yielded by the excitation wave travelling in distant areas of muscle. These are qualified by the adjective extrinsic.’

Thus it is apparent as Lewis²⁹ remarks elsewhere, ‘that we cannot be too circumspect in interpreting curves taken by leads direct from the heart muscle.’

Ultimate understanding of them would seem in the main to depend upon observing the effects of experimental change in the conditions under which such electrograms are taken, and it is only too apparent that the simple and somewhat clumsy experiment here reported would in normal circumstances represent one of a prolonged series of carefully controlled and repeated experiments. It will be noted that the application of cold to the surface to the right and left ventricles produced, as shown in c and d (fig. 3) very considerable changes in the deflexions of the electrograms taken from those leads, but the significance of these changes and their interpretation would appear to depend upon the carrying out of further experimental work.

Summary.

A case of ectopia cordis has been described. Electrocardiograms and electrograms were taken and the effects of cold on the electrograms recorded. It is felt that the interpretation of these results is dependent upon a further experimental investigation, but by virtue of the rarity of the conditions under which they were taken they appear to merit recording.

It is a pleasure to have this opportunity of thanking Dr. Hall for permission to publish details of this case admitted to his care, Professor Hume for the interest he has taken in it, and Dr. Carr for the assistance he gave in the taking of the electrograms.

REFERENCES.

1. Weese, C., *Inaug. Diss. Berolini*, 1819.
2. Torres, J. I. de, *Phil. Trans.*, London, 1739-41, XLI, 777.
3. Martinez, M., *Haller's Disputationes Anat. Select.*, Gottingen, 1747, II, 973.
4. Walter, J. G., *Mus. Anat.*, Berlin, 1805, I, 125.
5. Tourtelle, E., *J. de méd.*, Paris, 1784, LXII, 579.
6. Cruveilhier, J., *Gaz. méd. de Paris*, Paris, 1841, IX, 497.
7. Jones, S., *Trans. Path. Soc. London*, London, 1855, VI, 98.
8. Daniell, E., *Brit. Med. J.*, London, 1860, 776.
9. Schlesinger, H., *Berl. klin. Wchnschr.*, Berlin, 1870, VII, 376.
10. Barnardo, G. F., *J. Anat. & Phys.*, London, 1898, XXXII, 325.
11. Lilwall-Cormac, J., *Brit. Med. J.*, London, 1903, I, 1320.
12. Matteucci, *Zentralbl. f. inn. Med.*, Leipzig, 1904, XXV, 25.
13. Ellis, A. G., *Proc. Path. Soc., Philadelphia*, Philadelphia, 1906, IX, 36.
14. Greiffenberg, M., *Ztschr. f. Geburtsh. u. Gynäk.*, Stuttgart, 1908, LXII, 453.
15. Rolland, W., *Brit. Med. J.*, London, 1913, II, 121.
16. Cosgrove, S. A., and St. George, A. V., *Am. J. Dis. Child.*, Chicago, 1924, XXVII, 594.
17. Cutler, G. D., & Wilens, G., *ibid.*, 1925, XXX, 76.
18. Bloch, S., *Zentralbl. f. Gynäk.*, Leipzig, 1927, LI, 625.

19. Stiemens, M. J., *Nederl. Tijdschr. v. Geneesk.*, Amsterdam, 1929, I., 425.
20. Ledényi, J., *Anat. Anz.*, Jena, 1935, LXXIX, 277.
21. Barker, P., McLeod, A. G., & Alexander, J., *Am. Heart, J.*, St. Louis, 1930, V, 720.
22. Samojloff, A., *Arch. f. d. ges. Physiol.*, Berlin, 1910, CXXXV, 417.
23. Kountz, W. B., Prinzmetal, M., & Smith, J. R., *Am. Heart, J.*, St. Louis, 1935, X, 614.
24. *Idem*, *loc. cit.*, 605.
25. Craib, W. H., *Heart*, London, 1927-29, XIV, 71; *J. Physiol.*, London, 1928, LXVI, 49.
26. Woronzow, D. S., *Ztschr. f. Biol.*, Berlin, 1927, LXXXVI, 231.
27. Lewis, T., Oppenheimer, A., & Oppenheimer, B. S., *Heart*, London, 1919, II, 147.
28. Lewis, T., *The Heart Beat*, London, 1920, 63.
29. Lewis, T., Meakins, J., & White, P. D., *Phil. Trans. Royal Soc.*, London, 1914, B, CCV, 375.
30. Gatch, F., *Heart*, London, 1909-1910, I, 235.