TRICUSPID ATRESIA: A NEW RADIOSCOPIC SIGN

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The object of this paper is to call attention to a radioscopic sign recently observed in two cases of tricuspid atresia. To the best of the writer's knowledge, this has not been previously noted, and it would appear to be of value in establishing the diagnosis before more elaborate investigations are carried out.

The sign consists of the asynchronous pulsation of the anterior and posterior cardiac borders when viewed in the left (II) oblique position. In the normal heart, contraction is synchronous; the posterior border being formed by left ventricle and the anterior border by right ventricle, though occasionally, particularly if an insufficient degree of rotation is employed, the upper part of the anterior border may be formed by the right auricle. Whilst screening a child with cyanotic congenital heart disease it was observed that in the left (II) oblique position, the posterior cardiac border contracted appreciably after the anterior. At the time it was thought that this finding might have been due to either (1) left bundle branch block, or (2) replacement of the right ventricle as the main component of the anterior border by the right auricle, as would occur in tricuspid atresia when the right ventricle is non-functioning and hypoplastic. Since left bundle branch block had already been excluded by a full cardiogram, the latter hypothesis was accepted as the more likely. The diagnosis of tricuspid atresia was subsequently proved by angiocardiography.

Another child with tricuspid atresia and presystolic contraction of the anterior heart border has since come under observation.

In practice, the theoretical concept that a similar appearance might be produced by bundle branch block has not been borne out. Numerous cases of both left and right bundle branch block have since been screened and no asynchrony of ventricular contraction has been detected. Nor has this phenomenon been observed in any other condition with the exception of one case in which the diagnosis of tricuspid atresia although possible, was not proved.

CASE REPORTS

Case 1. Aged 8. Permanent cyanosis had been present from birth. The exercise tolerance was poor and squatting frequent. Deep cyanosis was present, the fingers and toes were clubbed, and the left ear was deformed. The height was 50 in. and the weight 42 lb. (average normal values 50.7 in. and 58.3 lb. (O'Brien et al., 1947). The blood pressure was 90/70. The heart was enlarged to the left and soft systolic murmurs were present at the apex and base. The second sound was slightly accentuated in the second left interspace. The hemoglobin was 162 per cent. A cardiogram (Fig. 1) showed abnormal left axis deviation (—40°). The precordial leads were unusual but suggestive of left ventricular enlargement. X-ray examination (Fig. 2) showed that the cardiac enlargement was due almost entirely to the left ventricle. The left auricle and right side of the heart appeared normal. On radioscopy in the left (II) oblique position the anterior border, which was of apparently normal contour was seen to contract appreciably before the posterior which was formed by the enlarged left ventricle. It is suggested that this was due to replacement of the right ventricle as the main component of the anterior border by the right auricle as the result of right auricular enlargement combined with hypoplasia of the right ventricle. It is interesting to note that the 387
FIG. 1.—Cardiograms of three cases of tricuspid atresia described in the text.

radiologist commented on the normal appearance of the right side of the heart in the left oblique view. Angiocardiography demonstrated a large communication between the right and left auricles, absence of filling of the right ventricle and filling of the aorta at 2½ seconds. These findings were considered diagnostic of tricuspid atresia. Operation was refused.

Case 2. Aged 8. Permanent cyanosis had been present from birth. The exercise tolerance was poor, minimal exertion producing severe dyspnœa. Deep cyanosis was present and the fingers and toes were clubbed. The height was 48 in. and the weight 40 lb. (average normal values 50-3 in. and 57-1 lb.). The blood pressure was 110/90. A harsh systolic murmur was audible over a fairly wide area, maximal over
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Fig. 2.—Postero-anterior and left (II) oblique radiographs of Cases 1, 2, and 3.
the sternum opposite the third and fourth interspaces. The haemoglobin was 134 per cent. A full cardio-
gram (Fig. 1) showed borderline abnormal left axis deviation (−30°) and left ventricular enlargement. The X-ray films (Fig. 2) revealed no radiological cardiac abnormality. On radioscopy, although the heart was normal in size and shape, presystolic pulsation of the right border was seen in the P-A view, and of the anterior border in the left oblique position. An angiocardiogram demonstrated a large communication between the right and left auricles, absence of filling of the right ventricle, and filling of the right sided aorta at 1½ seconds.

These findings were considered diagnostic of tricuspid atresia. In this case, prior to angiocardiography, the diagnosis was in doubt owing to the normality of the radiological appearances. The radioscopic findings, however, lent support to the diagnosis of tricuspid atresia.

A Blalock-Taussig operation was performed from which the patient made a good recovery. Owing to her condition at the time, however, exploration to verify the diagnosis was considered inadvisable.

The sign described has also been observed in the following case in which the diagnosis remains in doubt.

Case 3. Aged 14. Intense permanent cyanosis had been present from birth. The exercise tolerance was much limited by severe dyspnoea on minimal exertion. Deep cyanosis was present and the fingers and toes were clubbed. The height was 60-5 in. and the weight 79 lb. (average normal values 61-2 in. and 106-2 lb.). The blood pressure was 110/60. When first seen in 1947, a basal systolic thrill and a loud systolic murmur, maximal in the second and third left interspaces were present. Since then, this patient's condition has improved somewhat, while the thrill and murmur have become more pronounced. The haemoglobin was 138 per cent and a cardogram (Fig. 1) showed left ventricular enlargement with an electrical axis of 0°. X-ray films (Fig. 2) showed an unusual cardiac outline with an absence of projection of the heart border to the right. Nevertheless, the transverse diameter was at the upper limit of normal. In the left (II) oblique view, the left ventricle was seen to be enlarged; the anterior border of the heart showed no abnormality, but radioscopy revealed presystolic contraction of the anterior border. Angiocardiography demonstrated a large communication between the right and left auricles and absence of filling of the right ventricle.

The film at 2 seconds was unexposed and at 3 seconds both aorta and pulmonary artery were filled.

The evidence afforded was thus consistent with a diagnosis of tricuspid atresia, but this was not proved.

DISCUSSION

The principal signs on which the diagnosis of tricuspid atresia rests are permanent cyanosis associated with left axis deviation in the cardiogram, but the latter is not invariably present (Ash, 1950 and Taussig, 1947), nor is radiography always helpful, since the cardiac outline is not always typical and may even be normal. While, in the majority of patients, the diagnosis may be fairly straightforward, there are some in whom it is uncertain; it is suggested that the sign described above may be of value in establishing the diagnosis when angiocardiography and cardiac catheterization are not immediately practicable.

SUMMARY

What is believed to be a new sign of tricuspid atresia is described. This consists of presystolic pulsation of the anterior cardiac border in the left oblique position. Details of two established and one doubtful case of tricuspid atresia showing this sign are given.

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REFERENCES