RARE CARDIAC ANOMALY

COR TRILOCULARE BIVENTRICULAR IN MIRROR-PICTURE DEXTRO-CARDIA WITH PERSISTENT OMPHALO-MESENTERIC BAY,
RIGHT AORTIC ARCH AND PULMONARY ARTERY
FORMING DESCENDING AORTA *

BRET RATNER, M.D.

NEW YORK

MAUDE E. ABBOTT, M.D. AND W. W. BEATTIE, M.D.

MONTREAL, CANADA

In the remarkable case of congenital cardiac disease here recorded a complete absence of the interauricular septum is associated with other anomalies of a very unusual character, the whole forming a combination which, we believe, is unique in the literature.

CASE REPORT

Clinical Data.—A female infant, 17 days old, of Italian parentage was brought to the New York Nursery and Child's Hospital in a moribund state. The family history was negative. The child was of full term, normal delivery. Three days before admission the parents noted for the first time slight cyanosis of the extremities and somewhat difficult respiration.

On admission the infant presented extreme general cyanosis and Cheyne-Stokes' respiration. Both sclerae and conjunctivae contained extravasations, and several hemorrhagic spots were noted over the body. The apex beat was extremely faint—best heard to the right of the mid-clavicular line at about the sixth interspace. A faint prolonged systolic murmur was best heard at the base of the heart to the right of the sternum. The entire right thorax gave a flat note on percussion—no breath sounds were transmitted. The left thorax was dull on percussion—moist râles and harsh breathing were heard throughout. The heart outline could not be made out. The abdomen was distended and gave signs suggestive of fluid. There was a purulent discharge from the umbilicus. The liver was palpated below the umbilicus and the spleen 5 cm. below the ribs. The patient died two hours after admission. No blood culture was taken, and no thoracentesis was done.

Death was thought to be due to some form of congenital heart disease with cardiac failure, and also a possible septicemia and empyema.

Necropsy Summary.—At the necropsy the infection of the umbilicus was found to be superficial, and did not extend into the umbilical vein. There was marked engorgement of the abdominal contents and a small amount of free fluid in the peritoneal cavity. The liver, which was of normal size and markedly congested, was pushed down by a bulging right diaphragm. An interesting group of six supernumerary splenules were noted. Upon removal of the sternum, the pericardium was found to extend well out to the right, was greatly enlarged, and contained a moderate amount of transudate. The

---

*Received for publication July 16, 1921.

*From the pediatric Service of the New York Nursery and Child's Hospital (Dr. Oscar M. Schloss, Director) and the Department of Pediatrics, Cornell University Medical College, and the Medical Museum, McGill University.

*Read at the meeting of the American and Canadian Section of the International Association of Medical Museums at Cleveland, Ohio, March 24, 1921.

heart itself was tremendously enlarged and of a beef red color. The vessels and auricular portion were markedly distended with blood. The apex was broad and formed by both ventricles, giving the organ a globular shape. The right pleural cavity contained a large amount of transudal fluid—the left, a small amount. Both lungs were markedly engorged.

Description of the Heart.—Referring to Figure 1, it will be noted that there is a transposition of the ventricles and great vessels producing a “mirror picture” of these structures. The right chamber (left systemic ventricle) is somewhat smaller than the left chamber (right pulmonic ventricle), and on opening these chambers it was noted that the valve leading to the left chamber

ANTERIOR VIEW.

Figure 1

(pulmonary ventricle) was of tricuspid formation and the one leading to the right chamber (systemic ventricle) was of bicuspid formation. The ventricular septum is entire. The pulmonary artery arises from the anterior and right angle of the base of the pulmonic ventricle (left chamber) and runs upward and backward for about an inch to reach a plane posterior to the ascending aorta and terminates by dividing into three branches. A left and right pulmonary branch going to the hilum of each lung, the third branch, several millimeters long, a direct continuation of the parent stem, unites at an acute angle with what appears to be a corresponding branch from the ascending aorta. The latter is really the greatly narrowed continuation of the parent stem of the ascending aorta, the pulmonary artery forming the descending aorta through the greatly dilated ductus arteriosus.

The ascending aorta sprang from the base of the systemic ventricle (right chamber) and passed upward, forward and to the left. It extended for about one inch and also terminated by dividing into three branches, namely a left common carotid, a left subclavian, and a third short branch—a stenosis of the parent stem—which in common with the branch from the pulmonary artery described above (the dilated ductus arteriosus) gave rise to the descending aorta. These two anomalous short branches from the pulmonary artery and aorta were respectively found to be a patent ductus arteriosus and a stenosis of the isthmus aortae (Fig 1).

POSTERIOR VIEW.

Figure 2

The descending aorta immediately turned backward to descend along the right side of the vertebral column (right aortic arch).

Viewing the heart from behind (Fig. 2) the innominate artery is seen to arise from the descending aorta near its origin. The presence of a right aortic arch and the narrowing of the aorta at the isthmus are here seen more clearly.

Examination of the auricle reveals several features of great interest and rarity: (a) There is no trace of the interauricular septum, the common auricle opening below into the two ventricles through the two transposed auriculo-ventricular orifices; (b) there are four pulmonary veins (Fig. 2) disposed in

pairs on either side of the median line in the roof of the auricle, those entering on the right side being in close juxtaposition to the mitral orifice which leads into the right chamber (L. V.) and thence into the aorta, and those on the left in similar relation to the tricuspid orifice which leads into the left chamber (R. V.) and thence into the pulmonary artery; (c) the superior vena cava (Fig. 2) descends from above in its usual course but is continuous before its entrance to the auricle with an ovoid pouch-like sinus which lies vertically along the right lateral wall of the auricle and enters this by a narrow mouth which transmits the blood from the superior cava, no other vessel entering the sac. As stated above, this is thought to be a persistence of the dilated right omphalo-mesenteric vein at its entrance to the right sinus horn, the vein itself having become obliterated in the next stage of embryonic life. The inferior cava enters the auricle from below by a broad orifice with distinct lips which lies in such close relation to the opening of the superior cava through the sinus above described that its upper border forms the lower boundary of the latter (persistent double sinus venosus). The position of these structures on the right side of the common auricle is evidence that this chamber is not transposed in spite of the transposition of the ventricles and right aortic arch.

The ventricular walls, papillary muscles, and chordae tendineae are greatly hypertrophied.

Summary of Anatomic Findings.—Mirror picture transposition of ventricles and great trunks (true dextrocardia) and right aortic arch. Auricles not transposed but lie in normal relation to the reversed ventricles.

Complete absence of interauricular septum (cor trilobulare biventriculare) right pulmonary veins enter common auricle to right of median line. Superior vena cava entering persistent omphalo-mesenteric bay and inferior vena cava opening into auricle close to its mouth (double sinus venosus).

Ventricular septum entire. Patent ductus arteriosus and stenosis of the aorta at the isthmus, pulmonary artery forming descending aorta through greatly dilated patent ductus. Great hypertrophy of heart especially of pulmonic side. Multiple accessory spleens. Pleural effusion, great engorgement and passive congestion of viscera. Deep cyanosis (morbus caeruleus).

Diagnosis.—Beyond the existence of a dextrocardia it was, of course, impossible to distinguish during life the exact type of the complex cardiac anomaly here present, nor was a diagnosis attempted before death. The position, however, of the heart on the right side of the body without situs inversus of the remaining viscera pointed, as shown below, to an arrest of development at that period of early embryonic life when the heart was still situate to the right of the median line, before the speta had formed, such as might result in a bilobulate or trilobulate heart; and the marked cyanosis present would confirm this view.

The prolonged basal systolic murmur was undoubtedly due to the passage of the blood through the dilated ductus. A statistical study made by one of us (M.E.A.) has shown that the to and fro machinery murmur characteristic of patent ductus is a development of later life, and is not usually present at birth.

Pathologic Physiology.—Congenital cyanosis, or the symptom complex of deficient oxygenation of the blood through mechanical interference with the circulation by cardiac defects, may, as is well known, be produced by obstruction of the pulmonary circulation (by far the most serious cause and leading to the most extreme grades of cyanosis), or by mixture of venous with the arterial stream. In the present case, as intimated above, the latter cause was at work, for the pulmonary artery was widely dilated, and transmitted freely, both the blood to the lungs, as well as to the lower part of the body through the descend-

ing aorta, and the great hypertrophy of the pulmonic ("right") ventricle showed that these functions were being strenuously carried on.

The course of the blood may thus be traced briefly (Fig. 3): The blood from the superior and inferior cavae entering the right side of the common auricle meets the blood entering from the right and left pulmonary veins and passes synchronously through the transposed mitral and tricuspid orifices into the systemic and pulmonic ventricles whence it is distributed in a mixed stream to the head and upper extremities, a relatively small portion passing through the narrowed isthmus aortae to meet the current from the pulmonary artery to go

SCHEMATIC DRAWING SHOWING COURSE OF BLOOD.

Figure 3

into the descending thoracic and abdominal aorta supplying the lower parts of the body.

It is thus readily seen that under the anatomic conditions that existed, if the ductus had become closed the amount of aerated blood in the systemic circulation would have been very small, for the venous stream entering the aorta is relieved only by the aerated blood from the right pulmonary veins, while that entering the pulmonic ventricle from the left pulmonary veins would have been returned to the lungs and there performed an almost useless cycle. It is clear that both the position of the right pulmonary veins just above the right auriculo-ventricular orifice, and the condition of the "pulmonary artery becomes descending aorta" were conservative adaptations whereby all the aerated

blood available was distributed respectively to the upper and the lower parts of the body through the two great channels thus made available.

DISCUSSION

The peculiarities of the case lie, first, in the very early stage at which arrest of development of the structures entering into the formation of the auricle and auricular septum took place, as evidenced by the primitive mode of entrance of the superior cava through what is undoubtedly, as was pointed out to us by Prof. G. S. Huntington, a persistence of the dilated terminal end of the right omphalo-mesenteric vein of the early vitelline circulation, and the situation of the entrance of this in close juxtaposition to the entrance of the inferior vena cava (persistent double sinus venosus?); and the displacement of the right pulmonary veins to the right of the common auricle. Secondly, the curious and bizarre character of the dextrocardia, in that, in the presence of a "mirror-picture" transposition of the ventricles and great arterial trunks the auricular part of the heart remained untransposed, the great systemic veins entering a "persistent omphalo-mesenteric bay" lying on the right side of the common auricle. We know of no set of conditions identical with these in the literature, although analogous observations have been published by Hickman,¹ Abbott,² and recently Keith,³ in which transposition of the viscera (*situs inversus*) and of the auricles of the heart existed without transposition of the ventricles. The significant title "A Potentially Bicameral Heart" has been given by Dr. Keith to his recent very complete communication on this subject.

Other features, although by no means unique here, are the anomalous conditions of the systemic circulation, in that the main blood supply to the lower part of the body was transmitted to the descending aorta from the pulmonary artery through a greatly dilated ductus arteriosus. The main stream from the ascending aorta was directed through the great vessels of the neck to the upper part of the body, only a small portion passing through the stenosed isthmus to join the blood from the ductus arteriosus to enter the descending aorta. This condition of "pulmonary artery forming descending aorta" is usually compensatory in character, an adaptation to the needs of an obstructed or deoxygenated circulation from the left ventricle. Six such cases have been collected by one of us (M.E.A.) from the literature, and this is undoubtedly the explanation in the present instance. In a few rare instances the isthmus of the aorta is obliterated, or entirely absent, the ascending arch ending in the left subclavian while the pulmonary artery, dilated ductus, and descending aorta form a single large continuous

1. Hickman: *Tr. Path. Soc. Lond.* **20**:88, 1869.

2. Abbott, M. E.: *Osler's Modern Medicine*, **4**:345.

3. Keith: *Case of Transposition of the Viscera Showing a Potentially Bicameral Heart*, *Proc. Roy. Soc. Med. Lond.* **14**:3, 1921.

trunk; here a true anomaly existed due to the obliteration of the fourth, and the persistence of the fifth left primitive aortic arch. Such cases are reported by Hicks⁴ and Barlow.⁵

The late appearance of the cyanosis, which was first noticed on the fourteenth day, and became very deep on the seventeenth (day of death) is usual in all except the most extreme grades of interference with aeration such as pulmonary atresia when morbus caeruleus is present at birth; and it is quite characteristic of the more moderate mechanical causes at work in the present case, in which there was no pulmonary obstruction, but only admixture of streams.

The Dextrocardia.—As has been pointed out by Kuchenmeister⁶ and Nägeli⁷ in their masterly monographs on this subject, congenital dextrocardia in which the heart lies to the right of the median line and the apex points to the right, is of two types; in the one, a mirror picture transposition of the heart exists in which all parts may be reversed and may bear their normal relations to each other. This condition does not necessarily constitute an anomaly, as in such cases the mirror picture transposition of the heart is nearly always associated with transposition of the viscera. In the other type of dextrocardia the heart is not reversed on itself, but remains in the right side of the thorax, through an arrest of development at this stage; here the chambers are not reversed on themselves, and grave anomalies are always present owing to the early period at which this arrest occurs. The two types of cases may be differentiated by the electrocardiogram, the mirror-picture type giving a reversal of the normal electrocardiogram while the second type shows the usual "right ventricular preponderance" of a severe congenital cardiac lesion.⁸

The present complicated case is of the mirror picture type, and the complete development of the ventricular septum suggests that normal (although transposed) conditions originally prevailed here. It seems possible that the hitch lay in the failure of the vitelline circulation to become duly obliterated so that the right horn of the sinus venosus became fixed in its attachment on the right side, and prevented or nullified the transposition of the auricular part of the heart, the other changes being secondary adaptations. In any case the combination of these two anomalies is suggestive and may possibly give a clue to the cause of situs inversus and the period when this occurs.

The Omphalo-mesenteric Bay and "Double Sinus Venosus."—The distinct pouch in which the superior vena cava ends was considered by

4. Hicks: Malformation of Aorta and Pulmonary Artery, Tr. Path. Soc. Lond. **15**:85, 1864.

5. Barlow: Congenital Heart Disease, Tr. Path. Soc. Lond. **27**:141, 1876.

6. Kuchenmeister: Leipzig, Thesis, 1883.

7. Nägeli: Deutsch. Arch. f. klin. Med. **96**:552, 1909.

8. Lewis: Clinical Electrocardiography, London, 1913.

us at first a persistent sinus venosus in spite of the fact that no other veins entered it with the superior cava. Our thanks are due to Prof. G. S. Huntington for kindly giving us the correct explanation⁹ which is explained by Evans in his article there⁹ in the following words: "The vitelline veins empty their blood directly into the liver sinusoids, the blood from the left omphalo-mesenteric vein being collected by a short trunk which enters the left horn of the sinus venosus (v. hep. sinistra); but the right and larger one possesses a wide passage through this organ to the right horn of the sinus venosus."

A careful study was made by one of us (W.W.B.) of the comparative anatomy of the sinus venosus in relation to its opening into the auricle. The condition of double sinus venosus seen in *Lepidosteus* is simulated here, except that the coronary sinus did not open in connection with the other two veins. In *Lepidosteus* its homologue, the left ductus Cuvieri opens in common with the unpaired liver vein (inferior vena cava.) Rose.¹⁰

SUMMARY

1. Congenital dextrocardia is of two types due to (a) mirror-picture transposition (situs inversus) and (b) arrest of development. These types are of very different clinical manifestations and significance and may usually be readily distinguished.¹¹

2. A case is recorded of complete absence of the interauricular septum (ventricular septum entire) in congenital dextrocardia of the mirror picture or situs inversus type in which the auricles have remained untransposed.

3. The absence of the auricular septum is associated with an anomalous entrance of the superior vena cava through a "persistent omphalomesenteric bay" which represents a structure persistent from the early vitalline circulation and which had probably been an etiological factor in the lack of transposition of the auricles and possibly of the nondevelopment of the auricular septum.

4. The displacement to the right of the right pulmonary veins and the distribution of the blood from the pulmonary ventricle to the descending aorta through the pulmonary artery are probably compensatory processes, to further the distribution of the available aerated blood to all parts of the organs and are not independent anomalies.

5. The cyanosis present is due to admixture of venous with arterial blood and not to pulmonary obstruction, and its late appearance here is explained on the ground of its more moderate character until compensation had failed and pulmonary congestion occurred.

9. Keibell and Mall, 2:608 (Fig. 418) 1912.

10. Rose: Morpholog. Jahrb., 1903.

11. Meakins and Abbott: On the Differentiation of two Forms of Congenital Dextrocardia, Bull. Internat. Assn. Med. Museums, 5: (June 1) 1915.