THE ELECTROCARDIOGRAM IN SINGLE VENTRICLE

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Few electrocardiograms in cases of single ventricle have been published. Brown (1950) found that right axis deviation was usual, although left axis deviation occurred in one of his cases and he found others with no axis deviation. In cases where pulmonary stenosis is an associated defect, Campbell, Reynolds, and Trounce (1953) concluded that analysis of the cardiogram failed to reveal any basic common pattern in addition to that of right ventricular hypertrophy: five of their cases showed right axis deviation and evidence of hypertrophy of the right side of the ventricle. Taussig (1960) found it subject to wide variation: the standard leads may show either right or left axis deviation, and there may be gross disparity between the findings in the standard and in the precordial leads. Wood (1956) said that normal Q waves were usually seen in the antero-lateral chest leads or in one of the left-sided unipolar limb leads, despite the absence of the interventricular septum; otherwise the graph is apt to show changes that might ordinarily be more easily attributed to clockwise or anticlockwise rotation. Neill and Brink (1955) reported 8 cases of single ventricle with left axis deviation, 7 of which had precordial leads; a pattern of left ventricular preponderance was present in 3, of right ventricular preponderance in 1, and of equiphasic QRS in V 1–5 in 3: the P waves were increased in amplitude in 4 of these cases. Keith, Rowe, and Vlad (1958) suggested three patterns that may give one a lead in single ventricle: (1) right ventricular hypertrophy with a QR or rsR in the right precordial leads, (2) the pattern of negative complexes across the præcordium, and (3) the presence of equiphasic complexes across the præcordium. Honey, Rushton, and Taylor (1960) described a case of single ventricle, with a rudimentary chamber associated with inverted transposition of the great vessels, that showed a prolonged P–R interval and P mitrale.

The object of this paper is to review and discuss the electrocardiograms of cases of single ventricle seen at Guy’s Hospital and at Southampton Chest Hospital. Our criteria for acceptance into the study were that they should not have mitral atresia, tricuspid atresia, or a bilocular heart.

SUBJECTS

Since the six cases of single ventricle reported earlier by Campbell et al. (1953), a further 16 have been seen at Guy’s Hospital, including that of Honey et al. (1960). Five others were seen at Southampton Chest Hospital. From Campbell et al.’s 6 cases, only Cases 1, 2, and 6 satisfied our criteria and are reviewed here together with Honey et al.’s one case. In the whole group of 24 patients, 15 were male and 9 female. The ages ranged from a few weeks to 30 years. The diagnosis was confirmed by necropsy in 14 (including 2 with catheterization and angiography also and 1 with the latter also); by angiography in 7 (including 5 with catheterization also and 2 with operation also); and by operation in 1 case. Ten had inverted transposition of the great vessels, i.e. the aorta anterior and to the left, and the pulmonary artery posterior and to the right (this description applies only to those with levocardia). Ten had crossed or complete transposition of the great vessels, i.e. the aorta anterior and to the right, and the pulmonary artery posterior and to the left (this also
Anatomy of the vessels. Two applies only to those with levocardia. The remaining four had a normal relation of the great vessels. Two of the patients had dextrocardia with situs inversus, one had isolated levocardia, and 15 had pulmonary stenosis.

Findings

The findings are summarized in the Table.

The Axis of the Heart. On studying our 24 patients, the axis of the heart showed considerable variation. Five patients had left axis deviation with a mean QRS vector of −30 to −120 degrees, 11 had right axis deviation with a mean frontal QRS vector of +90 to −120 degrees, 6 had a normal axis ranging from −30 to +90 degrees, and the remaining 2 had either extreme right or extreme left axis deviation according to appellation, the axis of the heart being about −120 degrees. The axis of the heart bore no relation at all to the anatomy of the great vessels. In the 10 patients with inverted transposition, 2 had left, 5 had right axis deviation, and 3 had a normal axis. In the 10 with complete transposition, 1 had left, 5 had right axis deviation, 3 had a normal axis, and the remaining 1 had an axis of −120 degrees. In the 4 with normal relation of the great vessels, 2 had left, 1 had right axis deviation, and the remaining 1 had an axis of −120 degrees.

The P Waves. P mitrale occurred in Case 10 (Honey et al.’s case). It is of interest to note that the left-to-right shunt in this case was 20 l. per min. and that the systemic arterial and pulmonary
arterial O₂ saturations were 96·5 and 95 per cent respectively. Eight cases had P pulmonale ranging from 4–7 mm. in height; 4 of these had inverted transposition, 3 had complete transposition, and in 1 the anatomy of the great vessels was normal. Necropsy was performed on 4 cases with P pulmonale and the right atrioventricular valve was not stenosed in any of them.

The Præcordial Leads. Ten patients had the pattern of right ventricular hypertrophy with tall R waves in V1–2 and deep S waves in V5–6; 7 of these had right axis deviation, 1 had a normal axis, and the remaining 2 had an axis of —120 degrees. Seven patients had the pattern of left ventricular hypertrophy with deep S waves in V1–2 and tall R waves in V5–6: 4 of these had left axis deviation and the remaining 3 had a normal axis. Two patients had entirely negative deflections in the præcordial leads. The remaining 5 had biphasic deflections which were suggestive of biventricular hypertrophy. Eight had Q waves in the præcordial leads: in 4 of these the Q waves were in V1, in 3 they were in V6, and in the remaining 1 they were in V1 and V6. In the group of 4 with Q waves in V1, 2 had the pattern of right ventricular hypertrophy, 1 had that of combined ventricular hypertrophy, and the remaining 1 had negative deflections across the chest. The 3 with Q waves in V6 had the pattern of left ventricular hypertrophy. The one patient with Q waves in V1 and V6 showed the pattern of right ventricular hypertrophy. No special præcordial pattern could be correlated with a particular anatomy of the great vessels, for in the group of 10 with inverted transposition, 2 showed a pattern of left ventricular hypertrophy, 3 a pattern of right ventricular hypertrophy, 3 that of biventricular hypertrophy, and the remaining 2 had negative deflections across the chest. Three of these with inverted transposition had Q waves in V1 and another had a Q wave in V6. In the group of 10 patients with complete transposition, 3 had the pattern of left ventricular hypertrophy, 6 had the pattern of right ventricular hypertrophy, and the remaining 1 had the pattern of biventricular hypertrophy. In this group, one patient had a Q wave in V1, one in V6, and another one in both V1 and V5. In the group of 4 patients with normal anatomy of the great vessels, 2 had the pattern of left, 1 that of right, and the remaining 1 had that of combined ventricular hypertrophy. One patient had Q waves in V5–6. Neither could any definite pattern be correlated with pulmonary stenosis. In the whole group of 24 patients, 15 had pulmonary stenosis. Of these, 10 showed the pattern of right ventricular hypertrophy, 4 had that of left ventricular hypertrophy, and the remaining one had negative deflections across the chest. In the group of 10 with the pattern of right ventricular hypertrophy, 7 had right axis deviation, 1 had a normal axis, and the remaining 2 had an axis of —120 degrees. In the group of 4 with the pattern of left ventricular hypertrophy, 3 had left axis deviation and 1 had a normal axis. There was right axis deviation in the one patient with negative deflections across the chest.

High Voltage. In the whole group of 24, high voltage complexes occurred in 8 patients. In 5 of these, high voltage occurred in both the præcordial and limb leads, in 2 in the chest leads only, and in the remaining 1 in the limb leads only. The maximum height or depth of each deflection exceeded 35 mm. in each of the 8 patients, and in one the S wave in V3 measured 90 mm. Two of the 8 showed very tall and tented T waves which were in the præcordial and limb leads of one and in the præcordial leads only in the other.

Heart Block. Complete heart block occurred in 3 patients and a prolonged P–R interval occurred in Honey et al.'s one. Each of these 4 had inverted transposition of the great vessels. Wolff-Parkinson-White syndrome occurred in 1 of the 3 with complete heart block. An interesting feature in these three with complete heart block was the instability of the conduction defect: sinus rhythm, prolonged P–R interval, 2:1 heart block, complete heart block, normal QRS complexes, and Wolff-Parkinson-White complexes may all be recorded in the same record. In Case 6, lead V4 showed sinus rhythm with prolonged P–R interval, while the rest of the record showed complete heart block (Fig. 1). In Case 2, the same record showed sinus rhythm in VF, 2:1 heart block in V2, and complete heart block in lead I (Fig. 2). Case 5 deserves special mention. A record taken in November 1953 (Fig. 3) showed a qR pattern in V1 but there was no conduction defect. He had angiography in 1961 at which he developed complete heart block. A record taken on May 31, 1961 showed complete heart block, but some of the QRS beats had changed their vector in the antero-
posterior and frontal planes and had developed delta waves. One taken five months later showed sinus rhythm and left Wolff-Parkinson-White syndrome (Fig. 4). Another taken a month later showed 2:1 block and left W.P.W. syndrome (Fig. 5). During all this time, the patient had been on steroids which were ineffective in relieving the heart block; but when they were stopped, he developed cardiac arrest which responded to external cardiac massage.

**DISCUSSION**

*The Axis of the Heart.* The axis of the heart bore no relation to the anatomy of the great vessels nor to the presence or absence of pulmonary stenosis.

*The P Waves.* In the group of 8 cases of single ventricle studied by Neill and Brink (1955), 4 had P pulmonale; in one of these tricuspid stenosis was found at necropsy. Post-mortem examination was performed on 4 of our patients with P pulmonale and the right atrio-ventricular valve was
Fig. 3.—Cardiogram of Case 5, showing sinus rhythm in 1953.

Fig. 4.—Rhythm strips from Case 5. (A) Sinus rhythm on February 12, 1959. (B) Complete heart block and one beat with a delta wave on May 31, 1961. (C) Sinus rhythm and all complexes show W.P.W., October 14, 1961. (D) 2:1 heart block and W.P.W. syndrome on November 4, 1961.
FIG. 5.—Cardiogram of Case 5 shows left W.P.W. syndrome.

not stenosed in any of them. Honey et al. (1960) thought that the cause of the left atrial P waves in cases of inverted transposition might be correlated with the presence of inversion of the A-V valves, when the left of these is frequently incompetent. This explanation, however, will not account for the P mitrale in their patient since he had only an ejection systolic murmur ending 0.04 sec. before the single second heart sound. Since the ventricular diastolic filling resistance to each atrium must be the same in cases of single ventricle, the occurrence of P mitrale in the absence of left A-V valve disease remains unexplained. Although the ventricular diastolic resistance to each atrium is the same in single ventricle, the stroke volumes of the two atria are often different. It seems, therefore, that P mitrale or P pulmonale in single ventricle is likely to be related to the amount of pulmonary or systemic flows respectively. Cases with low pulmonary vascular resistance and a large L–R shunt may have a P mitrale, whereas cases with severe pulmonary hypertension or severe pulmonary stenosis and a large R–L shunt may have a P pulmonale.

The Præcordial Leads. No special præcordial pattern could be correlated with a particular anatomy of the great vessels. The occurrence of two cases of inverted transposition with negative deflections across the chest does not support the suggestion of Keith et al. (1958) that negative deflections of the QRS across the chest occur when single ventricle exists without transposition of the great vessels. Neither could any definite pattern be correlated with pulmonary stenosis. This confirms the earlier observation of Campbell et al. (1953) that analysis of the electrocardiogram in cases of single ventricle with pulmonary stenosis failed to reveal any basic common pattern in addition to that of right ventricular hypertrophy. In our view the only præcordial pattern suggestive of single ventricle is that of negative deflections across the chest. This pattern, however, occurred only in 2 of our 24 cases.
High Voltage. The occurrence of high voltage in cyanotic congenital heart disease is suggestive of single ventricle: it occurred in 8 of our 24 cases.

Heart Block. The most interesting feature was the instability of the conduction defect in our 3 cases with complete heart block. All our 4 cases with partial or complete heart block had inverted transposition of the great vessels. The development of left W.P.W. syndrome, as opposed to right W.P.W. syndrome, may be related to the reversal of the conduction system as described by Walmsley (1931). The development of W.P.W. syndrome in this case may throw some light on the mechanism of its production in some others. Its aetiology here cannot be due to accelerated conduction from the right atrium, since it developed during the phase of complete A-V dissociation. A focus of increased irritability in the ventricle may be responsible in this case (Grant, 1957). An example of single ventricle and complete heart block was that reported by Carns, Ritchie, and Musser (1941): here again inverted transposition of the great vessels occurred. Not all cases of single ventricle and heart block, however, have inverted transposition. The case described by Wilson and Grant (1926) of single ventricle and 2:1 heart block had ordinary complete transposition of the great vessels. Fig. 24.4 of Keith et al. (1958) of a case of single ventricle without transposition of the great vessels shows complete heart block.

In 1931 Walmsley described the bundle of His as being on the right side of the ventricular septum in a case of corrected transposition of the great vessels. Anderson, Lillehei, and Lester (1957) thought that the longer and anomalous pattern envisioned by Walmsley was probably of importance regarding the aetiology of A-V block, because the conduction must be by way of the right atrium and left atrium, and then the ventricular septum. Whatever the explanation of heart block is, in cases of inverted transposition with a ventricular septum, i.e. corrected transposition, a longer pattern of the bundle of His cannot be postulated in cases of single ventricle with inverted transposition, since the ventricular septum is absent. Again the reason why heart block occurs in those cases of single ventricle with complete transposition or with normal relation of the great vessels is far from clear. In the case described by Wilson and Grant (1926) the fibrous body instead of encircling the A-V bundle, as in the normal heart, penetrated it and subdivided it into fine strands. According to Mall (1912) and the later report on his work by Wilson and Grant (1926), "the A-V bundle apparently represents the remnants of the ring of the atrial canal joining auricle and ventricle. In the human embryo this ring tissue is broken anteriorly and laterally in the formation of the mitral and tricuspid valves. The remnant holds a constant position just behind the posterior endocardial cushion immediately over the septum of the ventricle, a position that is marked first by the interventricular foramen and later by the membranous septum. In destruction of the lateral and anterior walls of the atrial canal, not only does endocardial thickening play a role, but so does the connective tissue of the epicardium. The latter forms a collar encircling the A-V groove and as the mitral and tricuspid valves develop, the epicardial connective tissue is drawn into them, thus completing the separation of the atrial and ventricular musculature. Posteriorly it encircles the A-V bundle so that this becomes lodged between the posterior endocardial cushion and the outer connective tissue." Wilson and Grant thought that the greater destruction of the atrial canal in their case resulted in the bundle of His being penetrated by fibrous tissue and hence the 2:1 heart block. A study of cases of single ventricle seen at Guy's Hospital and others makes one conclude that abnormalities of the A-V valves are common in cases of single ventricle. A single A-V valve, rudimentary or malformed valves, cleft mitral or tricuspid valves, inversion of the valves, or mitral or tricuspid stenosis were all encountered. Mitral or tricuspid atresia also occurs with single ventricle. If these anomalies of the A-V valves in single ventricle represent an abnormal destruction of the primitive atrial canal, it is surprising that infiltration of the A-V bundle by fibrous tissue and heart block is not more frequent. It is probable that more than one factor is responsible for the development and behaviour of heart block in single ventricle.

Pattern of Left Ventricular Dominance in Single Ventricle. It is well known that a pattern of left ventricular dominance in the cardiogram is suggestive of tricuspid atresia (Astley, Oldham, and Parsons, 1953; Scott, 1955; Taussig, 1960). Seven of our patients had the pattern of left ventricular
Fig. 6.—Cardiograms of Case 7 (A) and Case 24 (B) who were originally diagnosed as having tricuspid atresia because of left ventricular preponderance.
dominance, four of these with left axis deviation. Cases 2, 7, 16, and 24, were originally thought to have tricuspid atresia mainly on cardiographic findings alone (Fig. 6). Cardiac catheterization, angiography, operation, or necropsy established the diagnosis of single ventricle in all four. This confirms the earlier observation (Neill and Brink, 1955; Nadas, 1957) that single ventricle is an important member of the group of cyanotic congenital heart diseases with left ventricular dominance in the cardiogram.

SUMMARY

Twenty-four cases of single ventricle were seen at Guy's Hospital and Southampton Chest Hospital. The electrical axis of the heart, the P waves, and the precordial leads were analysed in relation to the associated lesions in single ventricle. The cause of P mitrale or P pulmonale in single ventricle is thought to be related to the stroke volume of the left atrium and right atrium respectively. High voltage and the pattern of negative deflections across the chest are thought to be suggestive of single ventricle. No special precordial pattern could be correlated with the anatomy of the great vessels or with the presence or absence of pulmonary stenosis. Four cases had heart block.

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REFERENCES