# NON - FUNCTIONING OR UNDERDEVELOPED RIGHT VENTRICLE WITH INTACT VENTRICULAR SEPTUM *

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There is a group of congenital malformations of the heart in which blood flow to the lungs from the right ventricle is either very small or not existent and in which the systemic venous return for the most part travels from the right atrium to the left atrium by way of a foramen ovale or atrial septal defect to the left ventricle and out of the aorta. Blood flow to the lungs is by a patent ductus arteriosus or collateral circulation.

This course of blood flow is caused by either a small (underdeveloped) right ventricle or a pulmonary valvular atresia. In the latter group a ductus must be present. In the defects with small right ventricle and pulmonary stenosis some blood apparently reaches the lungs from the right ventricle and life may be sustained without a ductus.

The cases will be divided into three groups :

Group 1. Underdevelopment of the right ventricle with pulmonary stenosis with or without patent ductus arteriosus.

Group 2. Underdevelopment of the right ventricle with pulmonary valvular atresia with patent ductus arteriosus.

Group 3. Pulmonary valvular atresia with patent ductus arteriosus without underdevelopment of the right ventricle.

Autopsy examinations of the heart, at Texas Children's Hospital, of twelve patients who died between the ages of six days and seven years, revealed the following unusual constellation of the congenital defects, best summarized as "underdeveloped right ventricle or nonfunctioning right ventricle" because the hypoplastic ventricle seems to be the fundamental abnormality in these disorders.

In these twelve cases, three of them belong to Group 1, three of them to Group 2 and six of them to Group 3.

* Paper presented at the Second Middle Eastern - Mediterranean Pediatric Congress, Ankara, 1961.

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As described above, there is always an associated patent foramen ovale or interatrial septal defect in these cases which provides a circulation compatible with life

In such cases, it seems that in the prenatal circulation blood returned via the venae cavae, entered the right atrium and passed via the foramen ovale to the left atrium then to the left ventricle and to the aorta. Because of the stenotic tricuspid valve, pulmonic stenosis and the underdevelopment of the right ventricle, little blood flow would be passed via this route to the pulmonary artery.

Cooley and coworkers [1] reported that tricuspid stenosis or atresia is always associated with a poorly functioning or non - functioning right ventricle. Taussig [2] pointed cut the considerable significance of the size of the interatrial septal defects in such cases. Taussig also found [3] that if the defect is small, increased venous pressure and right auricular enlargement occur. Also, since the cardiac output is limited by the size of the defect, is is probable that patients with a poor interauricular communication will not be improved by the creation of an artificial ductus arteriosus. [3] We can summarize this explanation: there is a direct correlation between the severity of the stenosis of the tricuspid valve and the volume of the systemic blood which must pass through the atrial defect. Also a direct correlation may be considered between the size of the defect and increased venous pressure, right auricular enlargement showing peaked P waves in EKG, and also cardiac output. In other words output is limited by the severity of the tricuspid stenosis.

A review of the cases of this type of malformation at Texas Children's Hospital showed that all had an intact ventricular septum.

Of our cases, nine were female and three were male. In Group 1 there were two females, one male; in Group 2 there were three females; in Group 3 there were four females and two males. Birth weight was greater than five pounds for all except two cases in Group 2.

In Group 1 only one and in Group 3 only two patients' growth was normal, the others showed retarded growth. One individual in Group 1, one in Group 2 and two in Group 3 had histories of frequent infection. Fatigability was noted in one patient in Group 1, in all in Group 2 and in one in Group 3. Cyanosis was present at birth or right after birth in almost every case. The latest onset of cyanosis was at eleven months in a case in Group 2.

In all groups all cases showed increasing intensity of cyanosis except for two cases with constant cyanosis in Group 1. There were

cyanotic spells in all cases except two, one was in Group 1 and one was in Group 2. One patient in Group 1, two patients in Group 2 and two patients in Group 3 had periods of unconsciousness.

Clubbing of the fingers was only seen in two cases which were in Group 2, their ages were two years and three years, three months, respectively.

Onset of the murmur was at birth in 90 per cent of the cases, and within the first week in the others. The murmur was found to be of help. The location of the murmur varied due to the malformation. The character of the murmur was usually harsh and systolic in duration. The cases associated with patent ductus arteriosus had continuous murmur. In Group 3 all cases had a continuous murmur, loudest at the pulmonic area. In Group 2 two cases had no murmur and in the other the murmur was a faint grade 1. In all the cases of Group 2 there was a loud grade 2-4 systolic murmur at the lower left sternal border.

The pulse was small in one case in Group 1 and in two cases in Group 2; one was wide, one was bound in Group 3. The others were normal.

The greatest enlargement of the liver was four centimeters below the right costal margin. The liver was enlarged in all except two (both were in Group 3) which had normal sized livers.

The highest point of hemoglobin was 24.3 Gm. and the lowest was 14.1 Gm. Hematocrit varied between 46 and 68 per cent.

Arterial oxygen saturation determinations on these cases, by ear oxymeter, revealed a range of 67 per cent to 87 per cent. With exercise (crying for 3-5 minutes) oxygen saturation dropped to from 35 to 76 per cent. Oxygen saturation dropped with exercise in all cases. After exercise, with the administration of oxygen, oxygen saturation rose to between 72 and 90 per cent.

In all cases in which we have EKG records, rhythms were sinus. The axis varied between plus 130 right and plus 30 balanced. In Group 1 the axes were balanced, in Group 2 one was balanced, others were right, in Group 3 two were balanced, all others were right. The position of the heart was almost vertical in every case. In Group 1 only one individual had right auricular enlargement; in Groups 2 and 3 all had right auricular enlargement except one individual in Group 3 who had dextrocardia and had no right auricular enlargement. In Group 1 all cases had left ventricular hypertrophy and one of them also had strain. In Group 2 two cases had left ventricular hypertrophy, one had combined hypertrophy. In Group 3 four cases had right ventricular hypertrophy, two had

left ventricular hypertrophy, one of them had dextrocardia. Only one patient each in Groups 2 and 3 had first degree A - V block.

Two cases had slight, one had moderate, enlargement of the heart in Group 1; one had a normal sized heart and two had slight enlargement in Group 2; one had a normal sized heart, two had moderate and two had slight enlargement in Group 3; the sixth child was not examined radiographically.

Angiocardiography was done in one patient in Group 1; one in Group 2; three in Group 3; one patient in Group 3 also had retrograde aortography.

From the observations reported by Cooley and his co - workers [1] it appears that tricuspid stenosis or atresia with hypoplasia of the right ventricle together constitute one of the more promising fields for application of angiocardiographic methods. The interpretation of the changes is not very difficult. The three principal features of these anomalies can be demonstrated by angiocardiography as :

1. existence of an interatrial septal defect,

2. visualization of the diminutive chamber of the right ventricle or its absence,

3. presence of a large left ventricle.

The value of angiocardiography is great in this field of malformations. Technical improvements which are under investigation will further increase its validity in the future. Electrocardiographic and fluoroscopic findings, too, are of course needed for the diagnosis of these anomalies and must not be neglected.

The surgical procedures used in the treatment of congenital pulmonary stenosis are beneficial. In Group 1, one patient had left Blalock - Taussig [4] anastomosis end to side. One had Potts [5] anastomosis. In Group 2 only one patient had right Blalock - Taussig anastomosis. In Group 3 two patients had left Blalock - Taussig anastomosis, one had Potts. One had superior vena cava to right pulmonary artery anastomosis.

Age of death ranged from six days to three years, 5 months, in Group 1; seven weeks to seven years in Group 2; six days to two years, 5 months, in Group 3.

*Summary*

From our observations it appeared that in any case of underdevelopment of the right ventricle, whatever the cause may be,

the shift of the axis of the ventricular potentials toward the left can be expected, whether left ventricular hypertrophy is present or not.[6] There is more shift of the axis to the left in the above mentioned Groups 1 and 2, than in Group 3.

Cyanosis is usually present from birth. The mechanism of formation of this congenital anomaly is not yet known.

The history, clinical, laboratory and pathological findings have been reviewed. In all cases the findings were confirmed by postmortem examinations.

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