TWO CASES OF AORTICO-VENTRICULAR FISTULÆ

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An acquired fistulous communication between the aorta and the right ventricle, caused by syphilitic infection and rupture of an aneurysmal dilation of the right sinus of Valsalva, has been reported by Tompkins (1941). Schuster (1937) made mention of a similar mycotic rupture. Mall (1912) attributed the giving way of the wall in this region to the presence of a congenital weakness. The anatomical disposition of the three sinuses of Valsalva in relation to the rest of the heart was defined by Micks (1940) in his report of unruptured aneurysms of all three sinuses. Rupture of any one of the three sinuses, right, left, or posterior could occur. When the right sinus which is associated with the right coronary artery ruptures, it does so either into the right ventricle or the right atrium. The left sinus, associated with the left coronary artery, is related to the pericardial cavity, and death ensues rapidly through acute compression of the heart. On the other hand, communication with the chambers of the heart or with the adjacent pulmonary artery is compatible with life.

We report here two cases of aortico-ventricular fistulae. In one, an obvious rupture, death did not ensue until fifteen months after the onset of symptoms. In the other, we suggest that the fistula was a primary or persistent congenital one, rather than the giving way of the aortic wall at a later stage of life. Morgan Jones and Langley (1949) in their survey reported the incidence of such congenital fistulae. An anomalous channel between the aorta and the right ventricle was described by Brown and Burnett (1949): and this was interpreted as an accessory coronary artery. A similar interpretation has been given for the communication described by Edwards et al. (1958).

Case 1. A man, aged 25 years, was admitted for dyspnoea and palpitation of one year's duration. He was well nourished and had been an active athlete till the onset of the symptoms. There was no cyanosis. The pulse was 82 a minute, regular, and of collapsing form; the blood pressure, 130/50. The apex beat was thrusting. A continuous thrill was felt over the precordium and a continuous machinery murmur was heard over the entire precordium, both being maximal over the fourth left intercostal space. The lungs were clinically normal. The liver and spleen were not palpable. There was no oedema around the ankles. The teleradiogram showed the left ventricle enlarged and the lungs plethoric. The electrocardiogram showed left ventricular dominance with clockwise rotation of the heart.

The patient decided to seek treatment in London and was discharged from our care before investigations could be completed. An operation was performed in 1958 in London and revealed a rupture of a dilated right sinus of Valsalva with, in addition, a ventricular septal defect. There was no evidence of syphilis or endocarditis. The patient died three days after operation.

Comment. This was a congenital aneurysm of the right sinus of Valsalva which ruptured suddenly at the age of 25 years. The aneurysm itself had produced no symptoms prior to its rupture. In fact the patient was an athlete. In the case reported by Micks (1940) the aneurysm involved each one of the three sinuses, none of which ruptured: he attributed the cause of death to the pressure of the right sinus interfering with the conducting system. The anatomy of this particular sinus indicates that it is closely applied to the upper
part of the interventricular septum. One wonders to what extent the lack of physical support due to the defect in the septal region contributed towards the dilatation and eventual rupture of the sinus (Fig. 1).

**Case 2.** A girl, aged 2 months, was referred to the Thoracic Unit for investigation of permanent cyanosis since birth. She was the fifth in the family, all the other children being normal. The mother had not suffered from any infectious disease during the pregnancy. During the last month of her life, the child had had repeated attacks of dyspnœa during which the cyanosis deepened.

On examination, there was cyanosis in the lips, tongue, fingers, and toes, though the extremities were warm. The pulse was regular and 120 a minute. The apex beat was felt in the fourth space in the mid-clavicular line. No palpable thrills were felt in the praecordium or neck. Both heart sounds were heard. The second sound in the pulmonary area was single. A high-pitched systolic murmur was heard in the second left intercostal space. The breath sounds were normal and no adventitious sounds were heard in the lungs. The liver and spleen were not palpable. The haemoglobin was 86 per cent, the white blood count 11,000 per cu. mm., with a normal differential count. Radiologically the lung fields were oligemic, and the heart moderately enlarged with the right atrium dilated. The ventricular portion had a characteristic shape with considerable increase in curvature and slight prominence in the supero-lateral segment indicating that this was formed by the left ventricle.

In the ward the patient had repeated attacks of dyspnœa with increased cyanosis. At this stage of the investigation the patient died.

**Necropsy.** The heart, great vessels, and lungs were examined. There were minute scattered subperi-cardial haemorrhages. The lungs were normal. The heart appeared somewhat larger than normal. The aorta was slightly dilated and no dextroposition was observed. The pulmonary trunk was atretic proximally, tapering to a fibrous attenuated cord that was loosely connected to the heart surface and easily detached. Distally from a point just below its bifurcation, this artery was patent and connected to the aortic arch by a patent ductus arteriosus. Considerable hypertrophy of the left ventricular wall was noted with moderate dilatation of its cavity. The right ventricle was small and hypoplastic. The infundibular portion of this ventricle was not formed and there was no pulmonary lumen in the vicinity of the atretic cord representing the pulmonary trunk. The atra were large and there was a large atrial septal defect admitting one finger. The mitral orifice was normal. At the tricuspid orifice the valves were well formed though the orifice itself was smaller than the mitral. From just below the opening of the right coronary artery in the right sinus of Valsalva, a small communication passed through the membranous part of the interventricular septum to open into the right ventricle (Fig. 2).

**Comment.** In the month before death the child had repeated attacks of dyspnœa accompanied by increased cyanosis. When compared with the previous case, a similar catastrophe suggests itself at first sight—namely the sudden giving way of the right sinus. There are, however, particular features that counter the possibility of a sudden rupture. Careful observation of the hypoplastic right ventricular cavity failed to demonstrate an occlusive blood clot within it. Evidently blood did circulate through the right ventricle, the only outlet being the small channel leading into the aorta. The venous blood from the systemic
circulation probably entered the right atrium and from this the bulk of it was shunted through the large atrial septal defect into the left atrium. From here it passed through the mitral orifice together with the blood returning from the lungs via the pulmonary veins. The mixed blood was then pumped by the left ventricle into the aorta, some of which circulated through the lungs via the persistent ductus. Some of the right
atrial blood was likely to have entered the hypoplastic right ventricle through the tricuspid orifice and left it via the only exit already mentioned. Functionally of course this latter circulation could hardly have been of any significance (Fig. 3).

The autopsy findings of such a fistula leading from the right ventricle into the aorta coupled with the observation that a circulation did occur through this hypoplastic ventricle leaves us no alternative but to infer that this fistula corresponded to a minute defect in the membranous interventricular septum. The right ventricular blood was channelled into the aorta through the defect. The left ventricular blood reached the aorta normally. Distally, the pulmonary arteries were patent and were supplied with blood through the persistent ductus arteriosus.

Of interest here is also the observation made by Taussig (1947) that "defective development of the right ventricle is usually associated with tricuspid atresia and the absence of the normal flow of blood from the
right auricle into the right ventricle.” In this particular case, however, the hypoplastic rightventricle permitted a flow of blood through it and the tricuspid orifice was well formed and patent, though smaller than the mitral one. The shunt in this case was from right to left unlike that in the cases reported by Abbott (1919) and Abbott and Weiss (1928) where it was from left to right. Indeed, in all cases where the condition is truly acquired and where the pulmonary artery is normal, the shunt must of necessity be from left to right due to high pressure in the aorta as compared with that in the right ventricle. In our case the fistula was so small that there was no interference with conduction, as indicated by the regular pulse.

**SUMMARY**

We report here two cases identified as aortico-ventricular fistulae. In both the region of the right sinus of Valsalva was involved.

In the first case there was a rupture and death ensued fifteen months after the onset of symptoms. No infective condition hastened the rupture. A ventricular septal defect was observed in this case at the operation. It is possible that the lack of support to the right aortic wall (Fig. 1) due to the above defect contributed towards the aneurysmal dilatation of that part of the wall and its eventual rupture. The patient had been an athlete who led an active and strenuous life.

The second case was identified as a fistula of congenital origin. Such fistulae have been reported and Morgan Jones and Langley (1949) record their incidence in a series of cases. In our case we are suggesting that the fistulous communication represents a defective formation of the membranous ventricular septum. This of course means that the blood is channelled directly to the neighbouring aortic orifice.

In this latter case we also point out the existence of a circulation through the hypoplastic right ventricle, contrary to the generally accepted view, with a right-to-left shunt.

**REFERENCES**