CASE REPORTS

TRUNCUS ARTERIOSUS AND A SINGLE VENTRICLE

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A truncus arteriosus in association with a single ventricle is very rare and Siddoway and Chernish (1952) found only 16 possible recorded cases. In few instances has the clinical picture been described, and in all the cause of death has remained obscure.

Case Report

R. S., a full-term male infant weighing 5 lb. 9 oz. was born in the Obstetric Department of the Mayday Hospital following a surgical induction of labour at 39½ weeks for a very mild pre-eclamptic toxæmia (B.P. 150/90 and a trace of albuminuria). Pregnancy and delivery were otherwise normal and the fetal heart sounds were regular during labour. The baby was covered in meconium but was otherwise normal and cried well. The placenta was small and infarcted.

The baby remained well and took its feeds normally until 42 hours after birth when cyanosis developed and there was dyspnoea on handling and feeding. There was a loud systolic murmur over the præcordium maximal in the pulmonary area but no thrill was palpable. The lungs were clear. A diagnosis of congenital heart disease—probably tetralogy of Fallot—was made.

On the fourth day after delivery, tube feeding was commenced and continuous oxygen was given into a tented cot. Dyspnoea and cyanosis became worse and it was thought that broncho-pneumonia was developing. Penicillin was given and an "aevair" aerosol was passed into the tent. Pitting œdema, periodic gasping respiration, and lowered body temperature developed and coarse râles were heard all over the chest with impaired percussion note at the lung bases. By the morning of the fifth day considerable improvement with disappearance of the œdema had occurred. However, a sudden attack of intermittent gasping respiration and deep cyanosis occurred, frequent loose motions were passed, and ten minutes later the child died.

Necropsy. Full-term male infant with cyanosed head and extremities. On opening the chest, intense pulmonary congestion was seen, but the heart was not enlarged, the transverse diameter being 3.8 cm. (Fig. 1). The right atrium was moderately enlarged, the left being rudimentary and receiving blood from the pulmonary veins but having no connection with the ventricle. A patent foramen ovale 2 mm. in diameter had a crescentic valvular fold at its lower margin. The tricuspid valve was normal and there was no evidence of a septum in the single ventricle (Fig. 2). A single arterial trunk 1 cm. in diameter gave off the pulmonary arteries, each 3 mm. in diameter, and continued as the aorta, giving off innominate, left carotid, and left subclavian branches. A three-cusp valve guarded its base but no coronary ostia were found.

A single coronary artery 2 mm. in diameter arose from the innominate artery and passed down behind the large trunk between it and its pericardial sheath to the base of the heart where it divided into a right coronary artery running down the atrioventricular groove to supply branches to the right atrium and anterior surface of the ventricle, and a left marginal artery running down the left margin of the ventricle to the apex and supplying branches to the posterior surface of the ventricle. On sectioning the ventricular wall no rudimentary chamber was found. The descending aorta was of normal size. There were no other abnormal findings.

This case thus follows exactly the pattern of a truncus arteriosus of the solitary pulmonary trunk type. Its interest lies in the presence of ante-mortem general œdema and post-mortem pulmonary congestion.

Family History. The mother was operated on as a baby for pyloric stenosis. She had had rubella, mumps, measles, chickenpox, scarlet fever and severe rickets as a child. She could remember no mild illness during the pregnancy, which was her fifth.

FIG. 1.—Heart from 5-day-old baby, showing a single trunk leaving the common ventricle. This trunk has been opened to show the orifices of the pulmonary arteries which are illustrated by white markers. (1) œsophagus; (2) trachea; (3) innominate artery; (4) transverse aorta; (5), (6), (7) valve cusps; (8) right atrium; (9) common ventricle.

FIG. 2.—The heart has been opened to show the large right atrium, the common ventricle and the truncus arteriosus with a three-cusped valve at its base leaving the ventricle. (1) Right atrium; (2) common ventricle; (3) truncus arteriosus; (4), (5), (6) valve cusps illustrated by markers; (7) marker in patent foramen ovale; (8) marker showing entry of superior vena cava; (9) transverse aorta (continuation of truncus arteriosus; (10), (11) reflected flaps of ventricular wall; (12), (13), (14) cusps of tricuspid valve; (15) œsophagus; (16) trachea.

Her first pregnancy ended in a stillbirth at 33 weeks with breech extraction of a male foetus having deformed feet and genitalia. Her second pregnancy and delivery were normal but the boy was operated on at three weeks for pyloric stenosis. The third pregnancy and delivery were normal. The fourth pregnancy yielded a stillbirth at 40 weeks. The mother had a sister who had three miscarriages (one at six months) and two normal children. There was no other relevant family history.

Discussion

Three different developmental varieties of a single arterial trunk with a single ventricle all having a common clinical picture have been described.

(1) True truncus arteriosus communis persistens in which one abnormally large vessel leaves the heart giving rise to both systemic and pulmonary circulations without evidence of a second atretic vessel. The coronary arteries arise from its base.

(2) Solitary aortic trunk in which the pulmonary blood supply comes from the ductus arteriosus or from a collateral bronchial vessel and in which there may be evidence of an atretic pulmonary artery.

(3) Solitary pulmonary trunk in which an atretic aorta may be obvious or represented by a single coronary vessel given off from the main vessel or one of its major branches as in the above case.

The history of the present case and the findings on necropsy are typical, few cases surviving more than 20 days.

The case reported by Mehta and Hewlett (1945) was unique in surviving 56 years before death occurred from congestive cardiac failure in association with chronic bronchitis and unique in having a functioning mitral valve.

The most serious defect would thus appear to be the absence of a satisfactory return of blood from the lungs through a functioning mitral valve and it is probable that the cause of death, which is so poorly understood, is pulmonary oedema as a result of obstruction to the return of blood from the lungs—in effect a functional mitral stenosis of extreme degree. The obstruction might result at the foramen ovale from gradual closure of the orifice or from increased pressure in the right atrium due to a mild functional tricuspid incompetence. The increased systemic load produced by the effort of feeding or by anoxia could produce such incompetence by dilatation of the ventricle, thus precipitating a vicious circle which would inevitably end by death.

The only other explanation as to the cause of death is that offered by Rodgers and Edwards (1951), namely that an intra-pulmonary pulmonary stenosis occurs resulting from the single ventricle ejectile force which sends blood to both the pulmonary and systemic circulations. This explanation seems unlikely in view of the pronounced pulmonary congestion found at autopsy in all cases. Furthermore, it does not explain the long survival in the case of Mehta and Hewlett which is unique in the existence of a functioning mitral valve, because the pulmonary arteries must have been subjected to the same single ventricle ejectile force.

In the case now described the presence of ante-mortem oedema suggests a rise in pressure on the right side of the heart which would inevitably block the left-to-right shunt through the patent foramen ovale and thus lead to total obstruction of the pulmonary circulation, pulmonary congestion, and death.

The presence of other congenital abnormalities in the relatives of the case under discussion and the high foetal mortality in this family suggests some fundamental disorder that is affecting the embryos at an early stage of development. The coincident appearance of other abnormalities in the same child in several reported cases would suggest that if the disorder is a genetic one rather than an environmental one it is not related to a specific organ.

Summary

The clinical picture and post-mortem findings in truncus arteriosus associated with a single ventricle are discussed with illustration of a case.

The interesting haemodynamics of these cases are considered and a rise of pressure in the right atrium consequent upon dilatation of the common ventricle to cope with an increased systemic load is postulated as a cause of arrest of the pulmonary circulation and death.

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