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Congenital Absence of Ductus Arteriosus

—An Autopsy Case—

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Isolated absence of the ductus arteriosus is extremely rere condition although the ductus arteriosus may be hypoplastic or aplastic in association with other aortic arch anomalies.

Authors described a case of isolated agenesis of the ductus arteriosus documented by postmortem examination of a newborn infant who died of pneumonia following operation for a large omphalocele. The heart showed ventricular septal defect. However, no other cardiovascular anomalies were associated in this case. There were three vessels that were taking off from the aorta consisted of the right brachiocephalic artery, left common carotid artery and left subclavian artery. The anteriorly located pulmonary artery was divided into the right and left pulmonary anteries. There was no connection of vessel between the pulmonary artery and the aorta.

Key Words: *Congenital heart disease, ductus arteriosus, omphalocele, aortic arch anomalies*

INTRODUCTION

The ductus arteriosus is a main pathway of oxygenated blood from maternal side during intrauterine fetal life, accounting for 58% of oxygenated blood. The abnormally persistent ductus arteriosus in postnatal life is a relatively common cardiovascular malformation. However, congenital agenesis fo ductus arteriosus of isolated type has seldom documented in the literature. An autopsy case of this congenital agenesis of ductus arteriosus is described in a newborn baby who was born with omphalocele

CASE REPORT

This 11-day old male was born to a 26 year old mother by normal full term spontaneous delivery. The birth weight was 3.2kg. He was found to have a large omphalocele on his anterior abdominal wall. The chest x-ray showed a moderate cardiomegaly (Fig. 1). On the 5th day of life an operation for the omphalocele was performed. During the postoperative care the patient's condition became progressively worse and he expired 2 days after.

Postmortem examination revealed a generally pale and cyanotic baby with hydrocele and postoperative status of omphalocele. The heart was boot shaped with wide transverse diameter. It was enlarged and weighed 22.5 gm (normal, 19 gm). No remnant of ductus arteriosus was found between the pulmonary artery and aorta. The atrial septum showed a patent foramen ovale and the ventricular septum showed a perimembrauous round defect measuring 0.6×0.4cm. The lungs were bilaterally bilobed with microscopic foci of bronchopneumonia. Abdominal cavity showed multifocal fibrous adhesion and malrotated intestine with common mesentery. Other organs did

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not show any significant anomalies.

DISCUSSION

Congenital absence of the ductus arteriosus was first described by Stenson in 1670. However, the great majority of the cases was found as a combined anomalies in association with other intra- or extra-cardiac anomalies. Bharati et al. (1975) noted absence of the ductus arteriosus in 14% of 106 autopsy specimens with pulmonary atresia. However, isolated agenesis of the ductus arteriosus is extremely rare. We could find only one reference on this (Hamilton et al., 1982).

At an early stage of fetal development in human the ductus arteriosus develops from the distal portion of the left 6th aortic arch. The 6th aortic arch also known as the pulmonary arch, gives off an important branch which grows toward the developing lung bud. (Langman, 1969). On the right side the proximal part becomes the proximal segment of the right pulmonary artery. The distal portion of this arch loses its connection with the dorsal aorta and disappears. Meanwhile on the left, the distal part persists

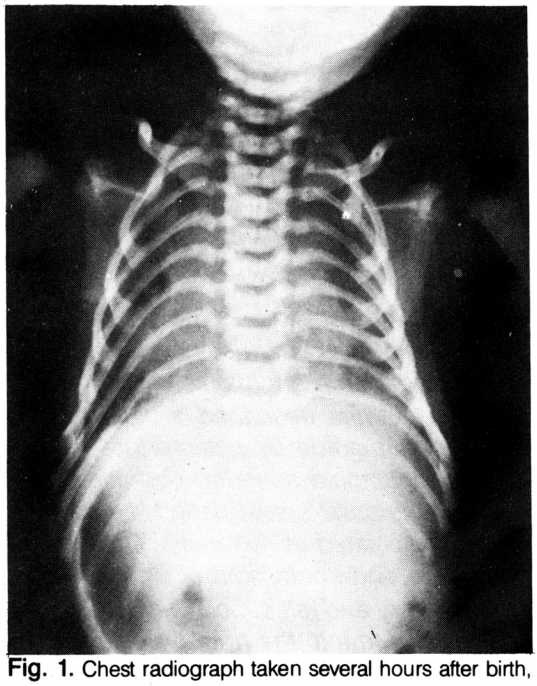

Fig. 1. Chest radiograph taken several hours after birth, showing a moderate cardiomegaly.

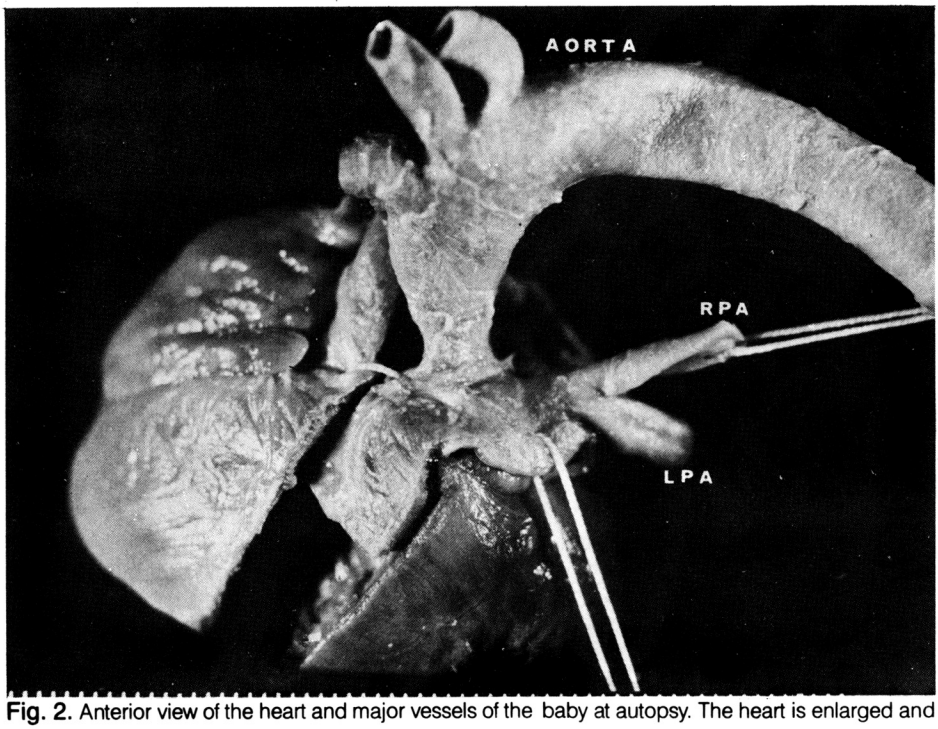

Fig. 2. Anterior view of the heart and major vessels of the baby at autopsy. The heart is enlarged and globular. Note the absence of ductus arteriosus.
RPA: Right pulmonary artery. LPA: Left pulmonary artery.

during intrauterine life as the ductus arteriosus. If on the left side, corresponding distal segment of pulmonary arch presumably loses connection with the left dorsal aorta, subsequently ductus arteriosus may not be formed.

By about the sixth week of gestation, the ductus arteriosus is sufficiently developed to carry the major proportion of the right ventricular output. Therefore the ductus arteriosus reduces the total work load of fetal ventricles. In present case the absence of ductus arterosus might have permited major proportion of the right ventricular output diverted through the ventricular septal defect to the left ventricle, leading to overload of the ventricles and accounted for cardiomegaly during intrauterine life. This condition can be comparable to the condition called premature closure of the ductus arteriosus which is a possible cause of intrauterine circulatory failure (Kohler, 1978).

The association of omphalocele and common mesentery in this case cannot be causally related to the cardiovascular malformation. However, these anomalies are involving the similar stage of development during the embryogenesis.

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