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Premature Closure of Foramen Ovale without Hypoplastic Left Heart Syndrome (An Autopsy Case)

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= Abstract =An autopsy case is described of a premature closure of the foramen ovale with a marked dilatation of the left atrium and an intact ventricular septum with aortic atresia but without left ventricular hypoplasia. This 28-week-old male baby was a stillborn to a 34-year-old multiparous mother. An intrauterine sonogram revealed marked ascites with anasarca, an enlarged left atrium and ventricle, a small right ventricle and breech presentation. An artificial delivery was done under the impression of congenital heart disease with hydrops fetalis.

The heart weighed 20 gm, and the left atrium and ventricle were dilated. The foramen ovale was completely closed with rudimentary fossa ovalis. Fibroelastosis was present in the right atrium and ventricle. Both lungs were trilobed and showed lymphangiectasia. It was presumed that in this case, the premature closure of the foramen ovale was the primary event, and the aortic atresia was secondary.

Key Words: ***Congenital Heart Disease, Atrial septum, Foramen ovale, Malformation, Pulmonary lymphangiectasia***

INTRODUCTION

The premature narrowing or closure of the foramen ovale is an infrequent but not rare anomaly. Analysis of 1,150 cases of congenital heart disease studied at the Congenital Heart Disease Research and Training Center revealed 10 examples of this anomaly in 1962 (Lev *et al.*, 1963). It may be associated with a hypoplastic left heart, mitral atresia, or stenosis, aortic atresia, or stenosis and congenital pulmonary lymphangiectasis.

Closure of the foramen ovale occurs after birth. The mechanism of the postnatal closure of the foramen ovale is circulatory change, that is, an increased pressure in the left heart (Moore, 1988). However, the mechanism of the premature closure of foramen ovale is unknown. Premature closure of the foramen ovale brings about hemodynamic changes in the heart and is associated with hypoplastic heart syndrome, mitral valve anomalies and aortic valve anomalies.

We have experienced an autopsy case of these anomalies that the closure of the foramen ovale was not associated with hypoplastic left heart syndrome. We present this case in view of its rare occurrence and also because of the unusual features associated with it.

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CASE REPORT

This male baby was a stillborn at 28 weeks of gestation to a 34-year-old multiparous mother. At six weeks of pregnancy, routine ultrasonography revealed a gestational sac of 2.2 cm in which a fetus of 0.2 cm with heart beat was present. The laboratory findings of the mother showed a hemoglobin level of 13.1 gm/100 ml with a hematocrit of 41.0%, and VDRL was non-reactive. At 12 weeks the fetal heart beat was present and the crown rump length was 5.4 cm.

However, the second follow-up sonogram at 27 weeks showed a breech presentation and a generalized edma with marked ascites. The left atrium and ventricle were enlarged. The thickness of the placenta had increased. At 28 weeks the pregnancy was interrupted under the impression of congenital heart disease with hydrops fetalis.

POSTMORTEM FINDINGS

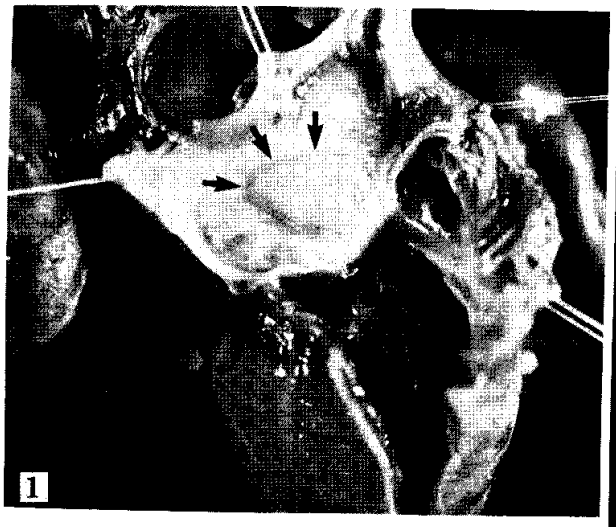
The baby weighed 1490 gm and the height was 35 cm. A marked generalized edema was present. The weight of the heart was 20 gm, and the great arteries were normally related. The superior vena cava and inferior vena cava were connected with the right atrium which was normally developed. However, when the right atrium was opened, there was a bulging of the atrial septum toward the right atrial side. There was no foramen ovale, of which the limbus of fossa ovalis could barely be identified (Fig. 1). The limbus was round-to-oval, and the maximum cross was 1 cm. Not a hint of probe-patency was present. The endocardium of the septum appeared white and slightly thickened. The tricuspid valve, right ventricle and pulmonary valve were normally developed. All pulmonary veins were connected with the left atrium, which was markedly dilated and measured 3 × 2.5 × 2 cm (Fig. 2). Its wall was tense and bulging in every directions. The pulmonary veins were also dilated. No evidence of collateral circulation was noted. The mitral valve showed a normal structure, but the left ventricle was slightly dilated(Fig. 3). The thickness of the left ventricle had increased and showed coarse trabeculations. The aortic valve was atretic (Fig. 4). Endocardial sclerosis was present in the left atrium and ventricle. The aorta was left-sided, and the ductus arteriosus was widely patent. Both lungs were trilobed and enlarged. The relationship between the pulmonary artery and the tracheobronchial trees was unremarkable. Microscopic examination of both lungs showed marked lymphangiectasia. Severe congestion was present in the liver and spleen. Acute tubular necrosis was present in the kidneys.

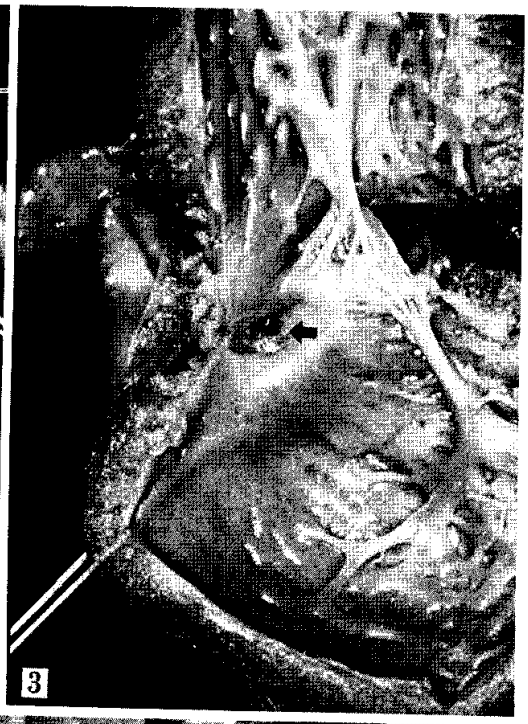
DISCUSSION

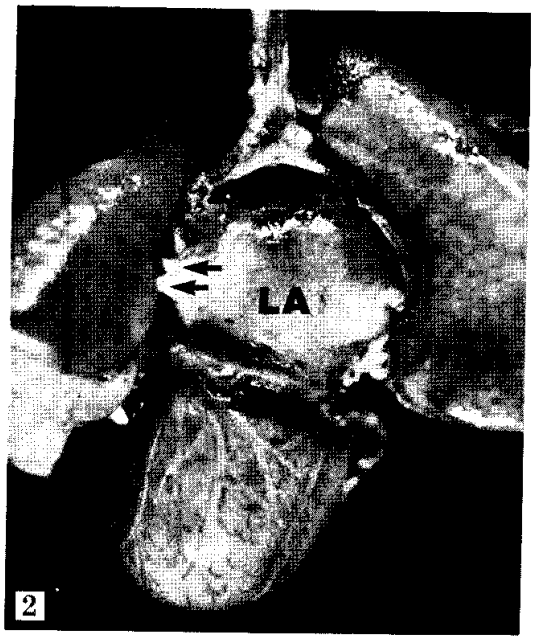
In late gestation, about one half of the fetal blood reaching the right atrium normally passes to the left atrium through the foramen ovale. When this flow is blocked by premature closure of the foramen ovale, the diverted blood presumably passes into the right ventricle, increasing the output of that chamber(Naeye & Blanc, 1964). From this point, blood must pass through the pulmonary circuit or through the still widely patent ductus arteriosus. However, in this case the outflow of the left ventricle is blocked by aortic atresia. Accordingly, an accessory pathway is essential for survival. Two collateral circulation paths have been known in this situation. The one is the levoatriocardinal vein and the other is the myocardiac sinusoid, through which blood flows in a retrograde fashion to enter the left circumflex artery where it is connected with the coronary sinus and, finally, the right atrium(Edwards & Dushane, 1950; Raghib *et al.*, 1965). However, these two alternate pathways could not be identified in this case. Therefore we should assume that some blood flow should have been present through the aortic valve until the time of the artificial termination of the fetus. Another possibility is that a small opening of microscopic size could have been present. In this case, the pulmonary lymphangiectasia most probably developed secondarily to the pulmonary venous obstruction.

The etiology of the premature closure of the foramen ovale is unknown. A possibility is suggested by Moerman *et al.*(1986). In hypoplastic left heart syndrome, because of the elevated left atrial pressure and a reversed interatrial pressure gradient, the valve of the forman ovale is opposed against the crista dividens and prematurely closed in the cases of the narrowing of the mitral and aortic valves. (Moerman *et al.*, 1986). In order to increase the pressure of the left atrium in the presence of an aortic atresia, mitral regurgitation must be present.

However, in this case the mitral valve was normally formed and mitral regurgitation was probably not present. From these points of view, this case is characterized by three unusual findings. First, despite the fact that an aortic atresia with an intact interventricular septum always accompanies a hypoplatic left heart, this case showed normal left heart. Second, although an aortic atresia blocked the outflow of the left heart completely, no evidence of collateral cir-







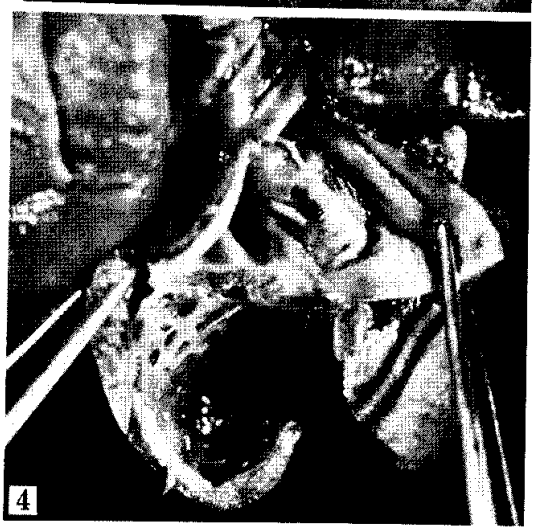


Fig. 1. The right atrium is opened to show the atrial septum that is completely closed. A hint of fossa ovalis is seen as a limbus(arrows). The endocardial surface is thickened with a whitish discoloration(endocardial sclerosis).

Fig. 2. Posterior view of the heart, showing a bulging left atrium(LA) and pulmonary veins(arrows). Note also the blunt cardiac apex. The lung shows lymphangiectasia on the pleural surface.

Fig. 3. The left ventricle is opened to show an atretic aortic valve(arrow) and a hypertrophic ventricular myocardium.

Fig. 4. The left atrium, tricuspid valve and left ventricle are well-developed. No evidence of hypoplastic left heart syndrome is present.

culation was present. Third, the size and location of the aortic arch was normal. These are the reasons why we believe that the premature closure of the foramen ovale in this case was most probably the primary event, and the aortic atresia was the subsequent phenomenon. At birth, with the presumed decrease in pulmonary resistance, there is an increase in pulmonary and left atrial and ventricular flow. The left side cannot accommodate this flow, which leads to pulmonary vascular enlargement, an increased right-to-left shunt through the ductus, right ventricular failure and death, This course of events proves to be the most lethal when a ventricular septal defect is not present, and is the least aggravating with a levoatriocardinal vein. Thus, death occurs in the first few days of life in most patients with these complexes, but survival may occur up to some months with a levoatriocardinal vein(Lev *et al.*, 1963)

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= 국문초록 =

좌심저형성을 동반하지 않은 난원공 조기폐쇄(1부검증례)

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저자들은 정상적인 심실중격과 대동맥 판막 폐쇄를 동반한 난원공 조기폐쇄부심 1례를 보고하다. 환아는 34세된 이머니에서 임신 28주에 태어난 사산아로 28주에 시행한 자궁내 초음파 검사에서 전신부종과 심한 복수가 발견되었고, 좌심방 좌심실은 커지 있었으며 우심방은 크기가 작아지 있었다. 임상적으로 태아부종을 동반한 선천성 심질환을 의심하여 인공 유도 분만을 시행하였다. 사후 검사에서 환아는 전신 부종이 있었고 심장의 무게는 20 gm이었고 좌심방, 좌심실은 확장되었고 난원공은 미발달된 난원와와 함께 완전히 닫혀져 있었다. 우심방 우심실에는 심유탄대증 외에는 정상적인 소견을 보였다. 양폭폐는 모두 3엽으로 구성되있고 크기는 조금 커서 있었고 림프관 확장을 동반하였다. 이 증례에서 저자들은 대동맥 판막폐쇄는 난원공의 조기폐쇄에 이어 2차적으로 발생되었다고 생각하였다.