

## Main Pulmonary Artery Stenosis Caused by Fibrocalcified Mass in a Young Infant

We present a rare case of main pulmonary artery stenosis secondary to protruding fibrous material in the main pulmonary artery associated with patent ductus arteriosus. A 1-month-old baby boy manifested cardiac murmur. Echocardiogram showed circumferential high echogenic mass inside the main pulmonary artery with pressure gradient of 49 mmHg and patent ductus arteriosus. The mass did not regress during 3 months' follow-up period. Angiographic images showed that the circular filling defect was located at the main pulmonary artery distal to pulmonary valve, and pulmonary valve and both pulmonary arteries were normal. After surgical removal of the circumferential material and ductus ligation, the pressure gradient became negligible. The material was consisted of scarcely cellular fibrous tissue, abundant coagulum of fibrinous material and dense calcification.

**Key Words :** Pulmonary Artery Stenosis; Ductus Arteriosus, Patent; Fibrous Tissue

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### INTRODUCTION

Pulmonary artery stenosis may develop anywhere along the pulmonary artery tree. The stenosis may be unilateral or bilateral, single or multiple. These stenoses are frequently associated with cardiac malformation, such as Fallot's tetralogy, complete transposition of the great arteries, pulmonary valve stenosis, persistent ductus arteriosus, ventricular septal defect or other complex malformation. The lesion may occur in isolation, but is quite rare in the neonate (1). Pulmonary artery stenosis secondary to fibrocalcified mass might be caused by other mechanism in comparison to usual pulmonary artery stenosis. We describe here a case of main pulmonary artery stenosis secondary to circumferential fibrocalcific material distal to pulmonic valve.

### CASE REPORT

A 1-month old baby boy was referred to our hospital for the evaluation of cardiac murmur. His birth weight was 3.7 kg, and there was no history of maternal illness during pregnancy. Maternal rubella antibody (IgM) was negative. The patient was alert active. Auscultation demonstrated grade 3/6 systolic murmur over the left sternal border. Except syndactyly between 3rd and 4th fingers of both hands, there was no anomaly. Chest radiography showed normal cardiac shadow and pulmonary vascularity. Electrocardiogram revealed right ven-

tricular hypertrophy. In echocardiogram, persistent ductus arteriosus was noted and a circumferential high echogenic mass inside of the main pulmonary artery was detected. The mass caused pulmonary artery stenosis with a pressure gradient of 49 mmHg (Fig. 1). After three months, there was no

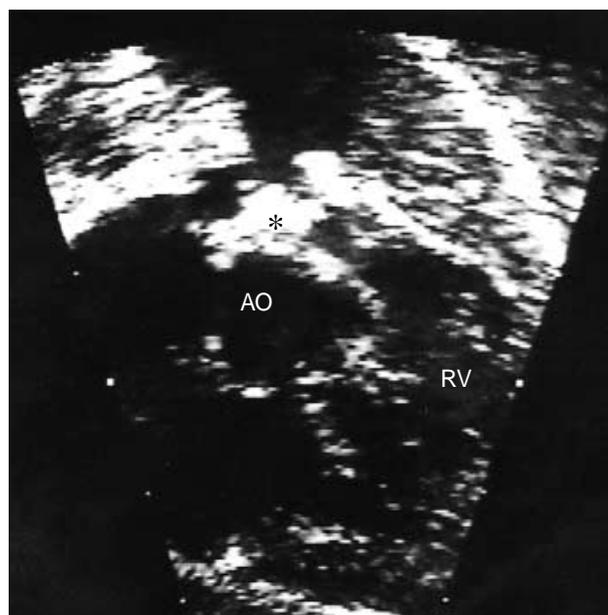


Fig. 1. Subcostal RAO view of pulmonary artery shows a high echogenic mass (asterisk) in the main pulmonary artery, causing pulmonary artery stenosis. AO, aorta; RV, right ventricle.

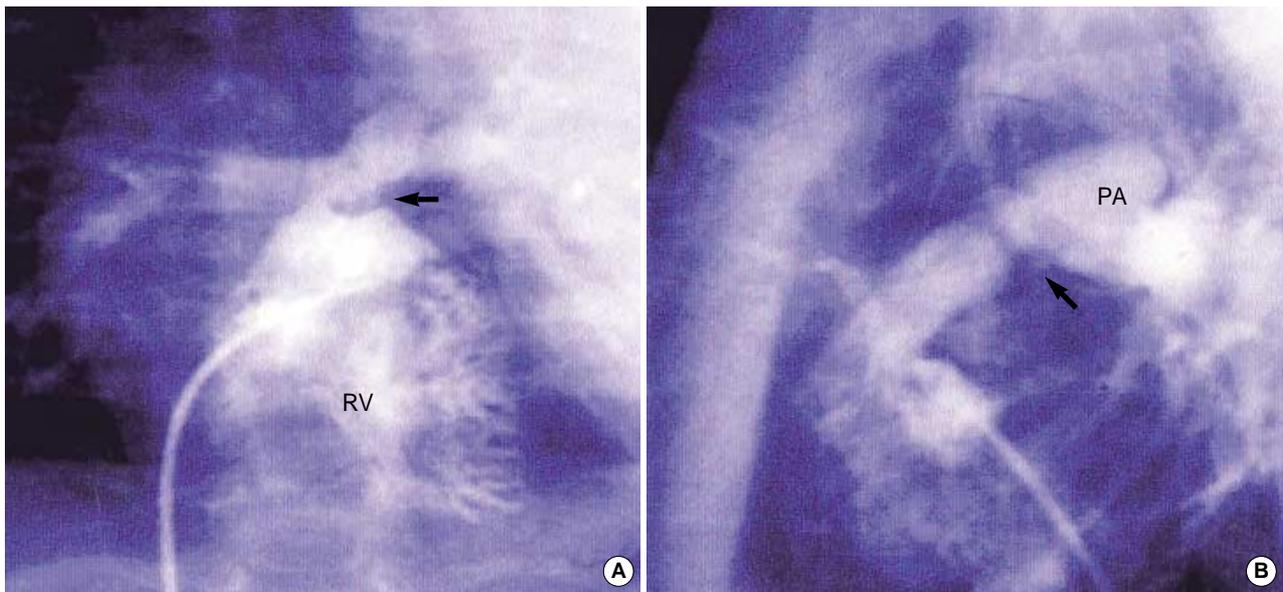


Fig. 2. Right ventricular outflow tract angiograms show a filling defect (closed arrows) inside the main pulmonary artery in antero-posterior (A) and lateral views (B). PA, pulmonary artery; RV, right ventricle.

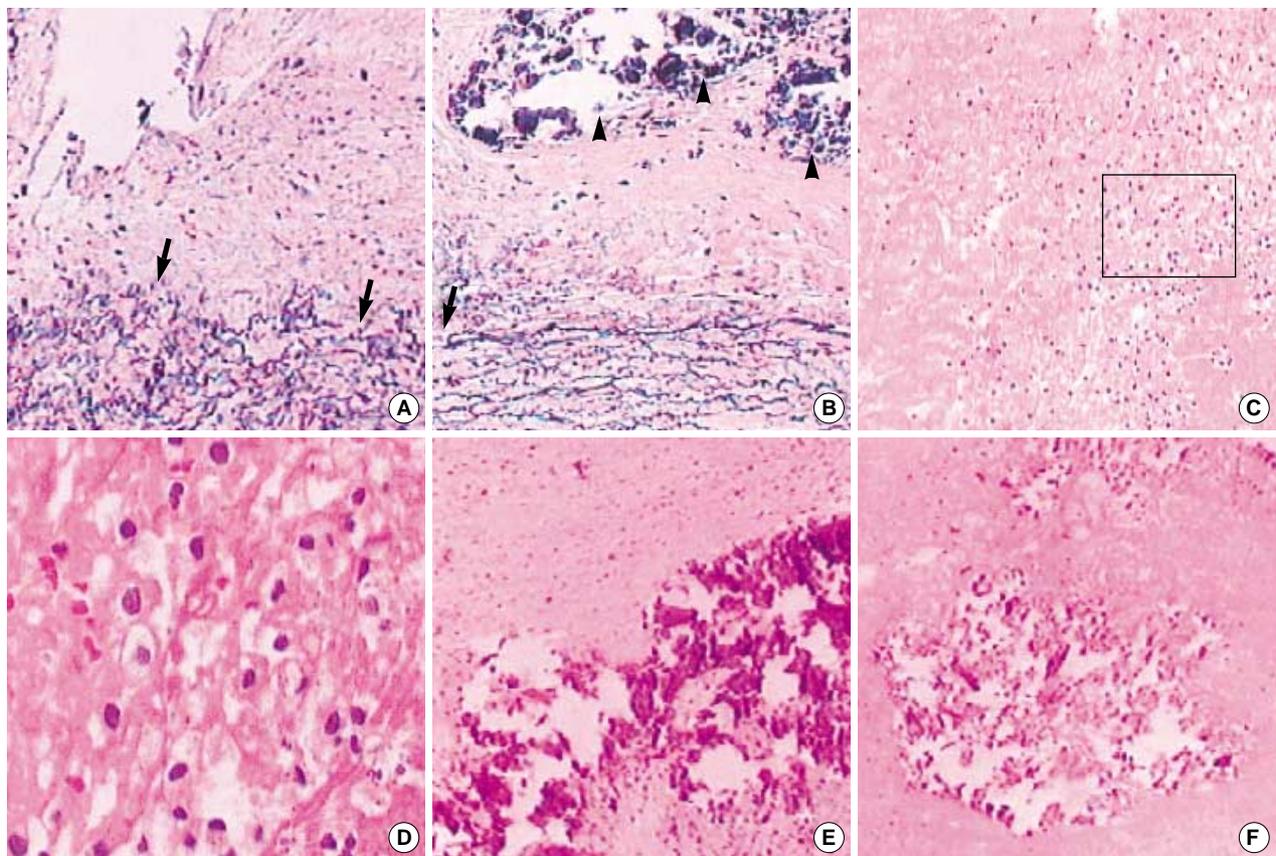


Fig. 3. Photomicrograph of the removed tissue from the pulmonary artery, consisting of intimal thickening by scarcely cellular fibrous tissue (A, B) and abundant coagulum of fibrinous material admixed with infiltrating macrophages (C, D). In thickened intima, irregularly-arranged, long spindle cells are admixed with a few mononuclear inflammatory cells (A, B). Destruction of internal elastic lamina (arrows in A and B) is present. Dense calcification is scattered in both cellular (arrowheads in B, E) and acellular areas (F). (A, B, elastic stain,  $\times 40$ ; C-F, H&E stain,  $\times 40$ ; D represents the boxed area of C,  $\times 2600$ ).

change in the mass size and pressure gradient. On cardiac catheterization, the pressure distal to the mass was 15/8 mmHg, and that of the right ventricle was 58/0 mmHg. At angiocardiogram, the right and left pulmonary arteries and the pulmonic valve were normal in size and shape. The circular filling defect in the main pulmonary artery distal to pulmonic valve was noted (Fig. 2). Under extracorporeal circulation, persistent ductus arteriosus was ligated and the circumferential mass lesion in the main pulmonary artery was totally removed. Histologically, the intima was markedly thickened by spindle cell proliferation, infiltration of mononuclear cells, and deposition of collagen. Most of the thickened intima underwent necrosis accompanying with advanced calcification. In and around necrotic coagulum of thickened intima, infiltrating macrophages were observed. Focal disruption of internal elastic lamina and degeneration of medial elastic tissues were also noted (Fig. 3). One month after the operation, the pressure gradient by Doppler method decreased to 13 mmHg and was negligible after two years.

## DISCUSSION

Pulmonary artery stenosis may develop as an isolated form in one third or in association with cardiac malformation, such as Fallot's tetralogy, complete transposition of the great arteries, pulmonary valve stenosis, persistent ductus arteriosus, ventricular septal defect, or other complex malformations. Luminal narrowing can occur anywhere from the pulmonary trunk to the segmental arteries (2).

Supravalvular pulmonary stenosis can be seen in Williams syndrome, Ehlers-Danlos syndrome, postrubellar syndrome, Noonan syndrome, Alagille syndrome, cutis laxa, or Silver syndrome (1). Our patient did not have any of these syndromes. Intrauterine viral myocarditis has been reported only rarely, however, infections with Coxsackie B virus (3), adenovirus (4), and parvovirus B 19 (5) have been occasionally documented. With the exception of rubella, the viral causation of significant valvular lesion in humans has received scanty support in the literature. Oyer *et al.* showed a case of fatal hydrops owing to adenoviral endomyocarditis with aortic and pulmonary valve stenosis, and myocardial fibrosis and calcification (4).

In O'Connor's series of supravalvular aortic stenosis (3 Williams syndrome, 2 familial, 1 sporadic), surgical or autopsy specimens showed microscopically disorganized medial element with fibrotic intima, sometimes containing lacunae. Ultrastructurally, thick irregular elastic fibers, abundant swirling collagen fibers, hypertrophied smooth muscles, and scant ground glass substance characterized the medial tissue defect (6). O'Connor also pointed out that, although hemodynamics during intrauterine development may predispose to localization of the stenosis to the supra-aortic valvar region, the cause for the mural dysplasia remains uncertain. In our case, the

specimen for histologic examination was only composed of mostly necrotic coagula, and small portions of intima and media. There was some evidence of inflammation and calcification in our case, but not in O'Connor's cases. Therefore, a direct comparison with O'Connor's cases was difficult.

In experimental animals, aortic intimal change as well as medial smooth-muscle cell hypertrophy, elastic fiber loss and fragmentation, and accumulation of excessive ground substance occur in response to experimental increase in intraluminal pressure (7). However, all of the deranged medial elastic pattern, the localized variation in the distribution and quantity of medial cellular element, the apparent deficient in the amount of ground substance, and the foci of extensive collagen deposition in space of the elastic network, are the findings that suggest abnormal prenatal development (6).

Description of the pathology of pulmonary stenosis following maternal rubella has been well appreciated. Histologically, intimal proliferation of the pulmonary arteries and dilatation of the distal branch have been reported. Occasional areas of thinning of the media were noted with fragmentation of elastic fibers (8, 9). In our case, histologically, the obvious fragmentation of the medial elastic tissue was not observed, but the intima was thickened by cellular proliferation and collagen deposition with a mild infiltration of mononuclear inflammatory cells to a variable extent. Also observed were the collagen degeneration and deposition of ground substances in the thickened intima, suggesting that the necrotic coagulum was the result of necrosis of thickened intima. Even though the sequence between intimal thickening and massive coagulation necrosis was not clear, the intimal proliferation might be responsible for the reduction in the lumen size. Although it has been suggested that a prenatal insult and histology of the lesion are not sufficient to support this picture in the neonate, all the clinical and histological features in our patient suggested that the hemodynamic changes with growth served as the stimuli upon the abnormalities, resulting in an obstruction of the developing pulmonary artery. Because the lesion was detected in the neonatal period, the contribution of some prenatal insult to development of pulmonary stenosis could be suggested. However, we could not prove it due to the lack of relevant history of congenital infection.

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