Tokai J Exp Clin Med., Vol. 2, No. 2, pp. 113–121, 1977

Pulmonary Artery Diverticulum Associated with Tetralogy of Fallot

Seiya MATSUYAMA, Taro WATANABE, Tsuneya WATABE
Yukio KURIBAYASHI, Takaaki TINA, Akira SHOHTSU*,
Mitsuru OSANO** and Tadashi INOUE***

*Department of Radiology and *Surgery, School of Medicine,*
Tokai University
***Department of Pediatrics and ***Surgery,*
School of Medicine,
Keio University

(Received February 12, 1977)

One hundred and ten cases of the angiographies of the tetralogy of Fallot were reviewed for the pulmonary diverticulum. It was found to be not unusual in association with the tetralogy, the frequency being 48% in the right aortic arch and 25.9% in the left aortic arch.

Frequent association with the right side aortic arch and with peripheral stenosis of the pulmonary artery suggested that the regression of the 4th as well as the 6th branchial artery plays an important role in the development of the diverticulum of the pulmonary artery.

Literature dealing with this subject was reviewed and only a few reports have been found to date.

(Key Words : Pulmonary Diverticulum, Blind Ductus, Tetralogy of Fallot, Right Aortic Arch)

INTRODUCTION

Right aortic arch and pulmonary artery hypoplasia are well-known associated anomalies of the tetralogy of Fallot (1,3,5,6,10,11). In 1961. Kerley et al. (7) described a small superficial bulge or a "blind ductus" of the pulmonary artery as an occasionally associated pulmonary artery anomaly in the tetralogy of Fallot or pulmonary stenosis. Since then, Hudson (1965) (2) and Ikeda et al. (1969) (4) reported this pulmonary artery anomaly and referred to it as a pulmonary artery diverticulum.

We recently reviewed 110 cases of the angiography of the tetralogy of Fallot which had been confirmed angiographically, surgically or by autopsy. We observed this phenomenon quite frequently in our own material and are convinced that this anomaly is not incidental but at least to some extent related to the anomalous development of the branchial arteries which is often associated with the tetralogy of Fallot.

We will report 34 cases of this anomaly in association with 110 cases of

Presented at the 33rd Meeting of the Japan Radiological Society, April 6, 1974
Department of Radiology and Surgery, School of Medicine, Tokai University, Bohseidai, Isehara, Kanagawa 259–11, Japan
Department of Pediatrics and Surgery, School of Medicine, Keio University, Shinanomachi, Shinjuku-ku, Tokyo 160, Japan.

the tetralogy of Fallot, 13 of which occurred in the tetralogy of Fallot with right-sided aortic arch.

MATERIALS AND METHODS

A total of 110 patients were examined by biplane serial right ventricular angiography for suspected tetralogy of Fallot or cyanotic heart disease. The angiographies were reviewed for a "diverticulum" or a "blind ductus" of the pulmonary artery, the location of pulmonary artery stenosis or right ventricular outflow obstruction and for the ratio of the caliber of the main pulmonary artery to that of the aorta (PA/AO) at the supravalvular level.

Of 110 patients, 50 were male and 60 were female (the age ranged from 2 months to 36 years) (Table 1)

Table 1 Number of cases of tetralogy of Fallot with or without pulmonary diverticulum

Age	With divericulum	Without diverticulum
0-5	26	49
6-10	4	13
11-15	2	6
16-20	2	4
21-	0	4
Total	34	76

Sample Cases

Case 1. Y.E. Female aged 10 months. Tetralogy of Fallot with left-side aortic arch (infundibular and valvular stenosis of the pulmonary artery) (Fig. 1)

Case 2. Y.U. Female aged 2 years. Tetralogy of Fallot with right-side aortic arch (infundibular stenosis of the pulmonary artery) (Fig. 2)

Case 3. K.Y. Female aged 4 years. Tetralogy of Fallot with left-side aortic arch (infundibular and peripheral stenosis of the pulmonary artery) (Fig. 3)

Case 4. H.U. Female aged 8 years. Tetralogy of Fallot with left-side aortic arch (valvular stenosis of the pulmonary artery) (Fig. 4)

RESULTS

Of 110 cases with the tetralogy of Fallot, 34 diverticula (30.9%) were observed in the proximal pulmonary arteries. There were many gradations of this abnormality varying from a small superficial bulge to a true "diverticulum". However, none of them was considered to be a true aneurysm. They included eight (23.5%) small pulmonary artery bulges and 26 (76.5%) diverticula. The number and the location of the pulmonary diverticulum and the correlation with the aortic arch in the tetralogy of Fallot are shown in Table 2. As shown in the table, 12 diverticula (48%) were found in the 25 cases of the tetralogy of Fallot with right aortic arch, six being located at the main pulmonary artery, four at the

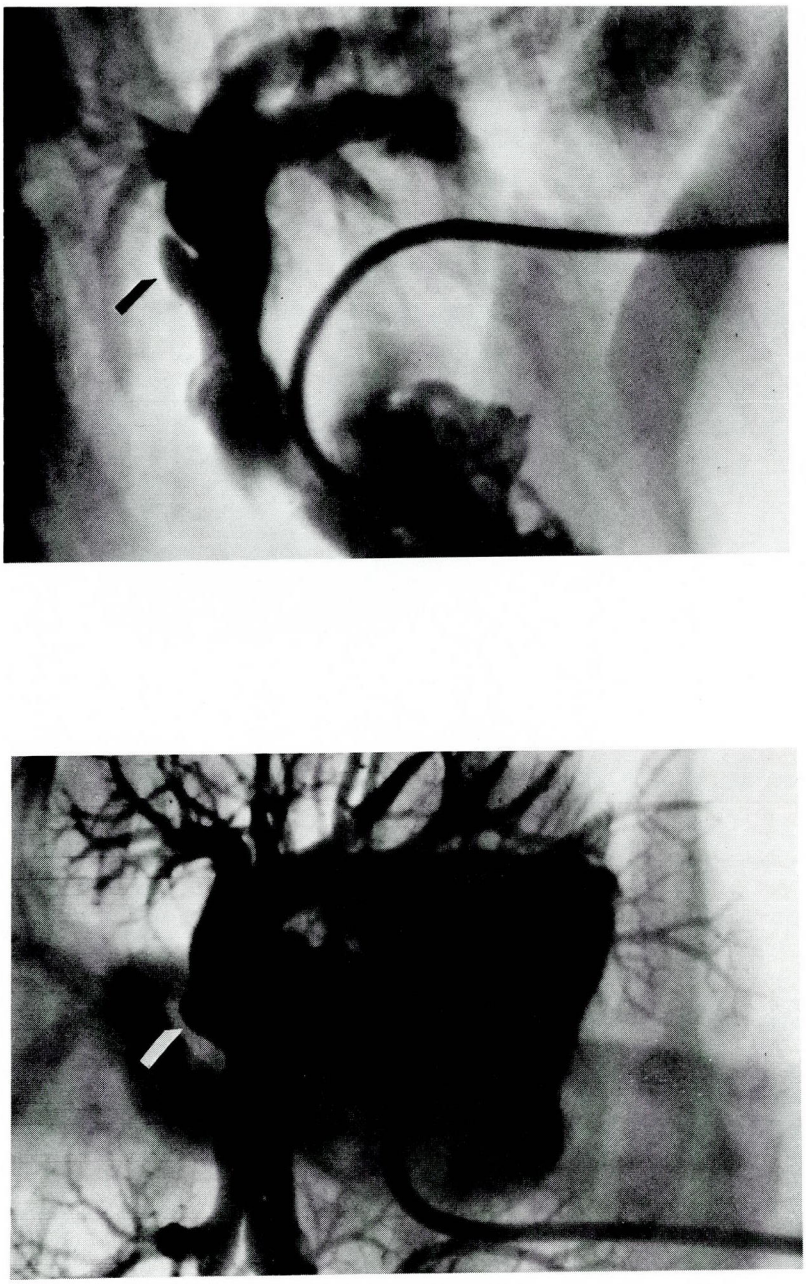

Fig. 1 Case 1. Pulmonary diverticulum (arrow) at the main pulmonary artery

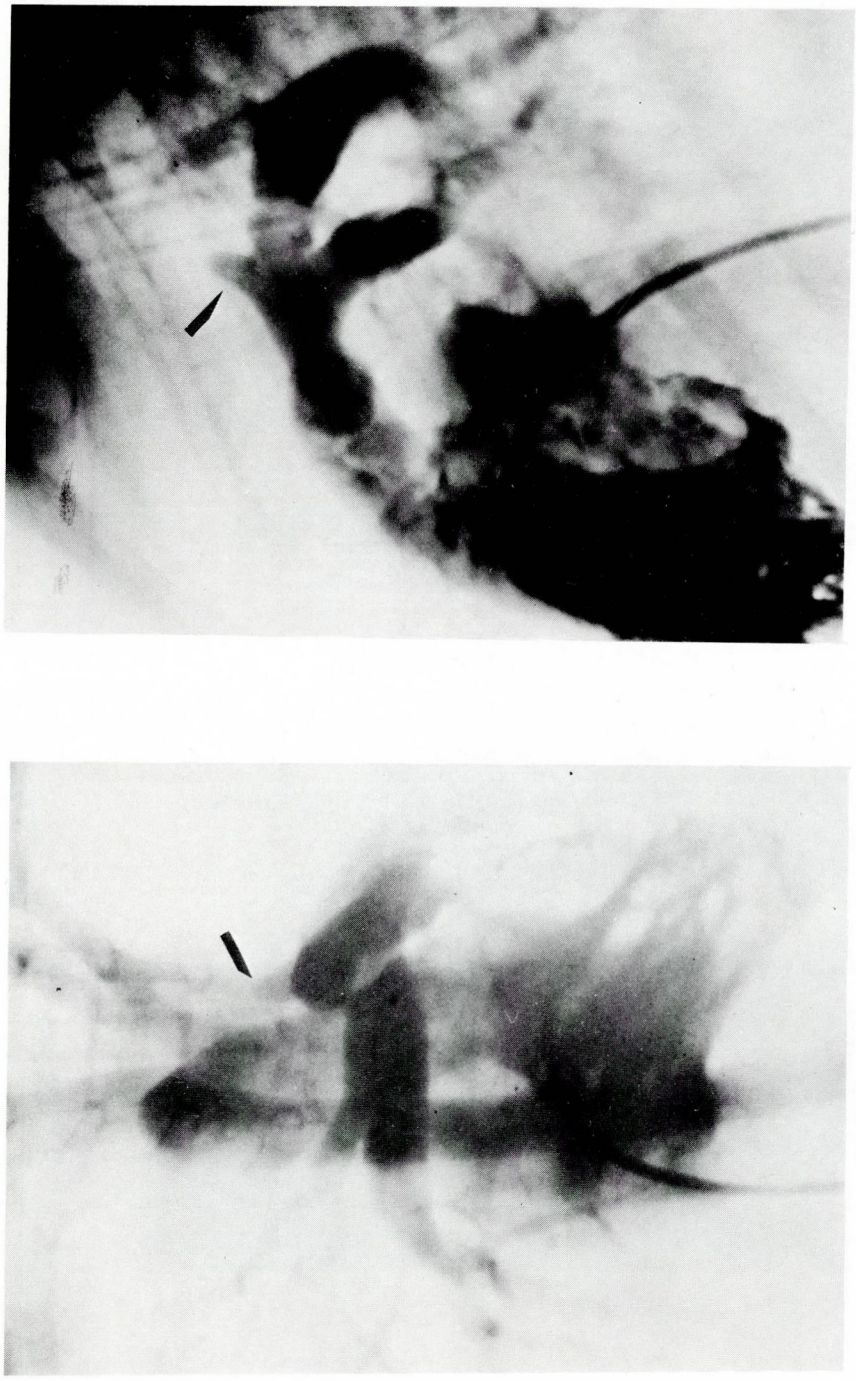

Fig. 2 Case 2. Pulmonary diverticulum (arrow) at the proximal left pulmonary artery

Fig. 3 Case 3. Pulmonary diverticulum (arrow) at the pulmonary artery bifurcation

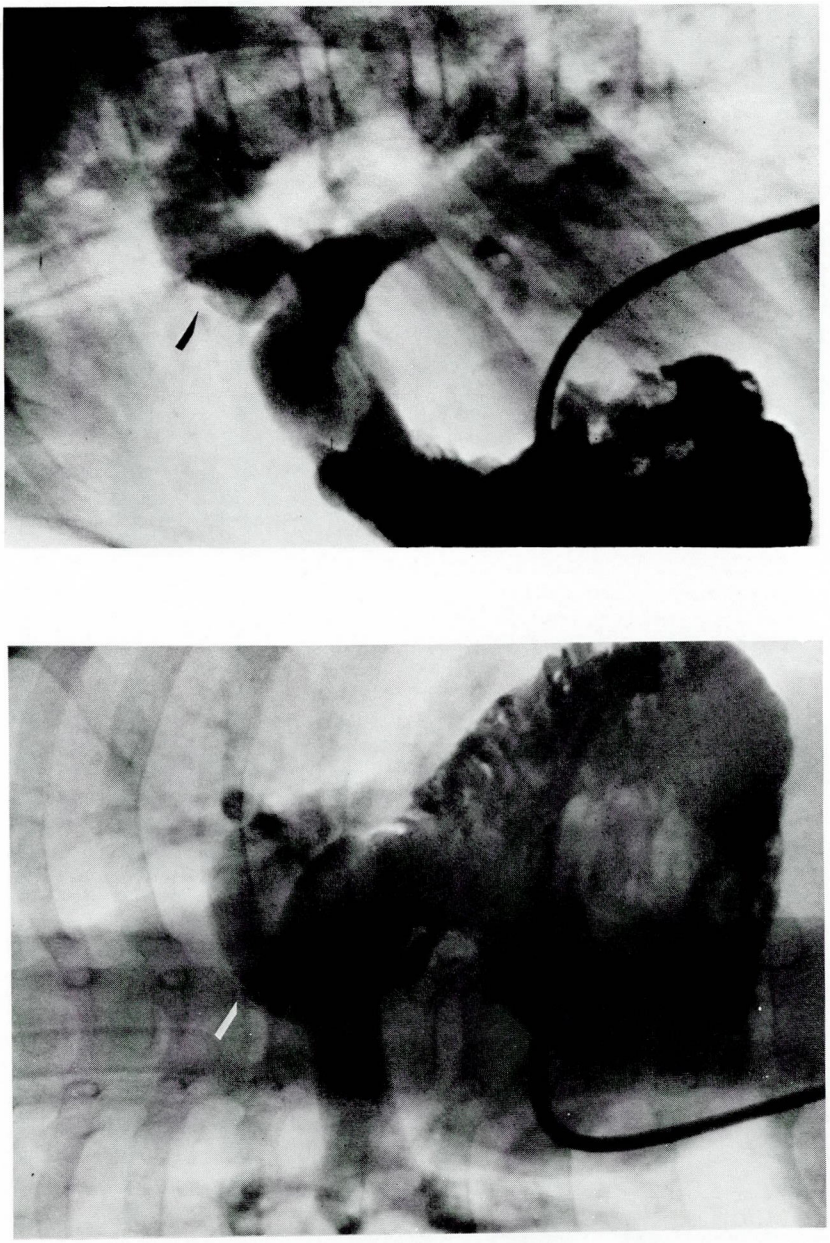

Fig. 4 Case 4. Pulmonary diverticulum (arrow) at the proximal left pulmonary artery

bifurcation and two at the proximal left pulmonary artery. Whereas, out of the 85 cases of the tetralogy of Fallot with left aortic arch, the diverticulum was found in 22 cases (25.9%), 10 of which were at the main pulmonary, four at the bifurcation and eight at the proximal left pulmonary artery. Patent ductus arteriosus was also found to be associated with two cases of the tetralogy with right aortic arch and one case of the tetrad with left aortic arch (Table 2). One case of the tetralogy with right aortic arch showed the diverticulum at the proximal innominate artery at the level of the ductus arteriosus (Fig. 2).

As for the location of the stenosis of the pulmonary artery or the right ventricle in the cases reviewed, 20 of 34 cases of tetralogy with a pulmonary artery diverticulum had both infundibular and valvular stenosis of the pulmonary artery. Supravalvular or peripheral pulmonary artery stenosis seems to be observed more frequently in the tetralogy of Fallot with a diverticulum (55.9%) than in those without (35.1%) (Table 3). Table 4 shows the relationship of the PA/AO ratio of the tetralogy of Fallot with diverticulum and that of the tetralogy without diverticulum. There was no significant correlation of PA/AO caliber ratio with the frequency of the pulmonary diverticulum (Table 4).

Table 2 Ductus arteriosus and variants in 110 cases of tetralogy of Fallot

Aortic Arch	PDA	Blind ductus				
		Pulmonary artery				Aorta
		MPA	Bifurcation	LPA	RPA	
R. Aortic Arch		6	4	2	0	
25	2		12			1
(22.7%)	(8.0%)		(48%)			(4.0%)
L. Aortic Arch		10	4	8	0	
85	1		22			0
(77.3%)	(1.2%)		(25.9%)			(0%)
Total						
110	3		34			1
(100%)	(2.7%)		(30.9%)			(0.9%)

Table 3 Pulmonary diverticulum in terms of type of pulmonary stenosis (tetralogy of Fallot)

Type of pulm, stenosis	With diverticulum No. of cases	Without diverticulum No. of cases
Infundibular (I)	31 (91.2%)	72 (97.3%)
Valvular (V)	23 (67.6%)	16 (21.6%)
Supravalvular (S)	19 (55.9%)	26 (35.1%)
(I) + (V)	20 (58.8%)	32 (43.2%)
(I) + (S)	17 (50.0%)	26 (35.1%)
(S) + (V)	12 (35.3%)	14 (18.9%)

Table 4 Pa/Ao ratio and pulmonary diverticulum (tetralogy of Fallot)

Pa/Ao	With diverticulum No. of cases	Without diverticulum No. of cases
0-0.3	1 (3.0%)	0
0.3-0.5	13 (38.2%)	23 (32.4%)
0.5-0.7	10 (29.4%)	28 (39.4%)
0.7-	10 (29.4%)	20 (28.2%)
Total	34 (100%)	71 (100%)

DISCUSSION

Kerley et al.(7) reported four cases of pulmonary artery diverticulum associated with the tetralogy of Fallot or pulmonary artery stenosis and referred to as a blind ductus arteriosus with the closure of the aortic side of the ductus. In 1959, Rudhe et al.(9) described a shallow bulge at the point of the anterior wall of the main pulmonary artery well above the pulmonary valve usually at the point of bifurcation on angiograms of the cases of pulmonary stenosis and the tetralogy of Fallot. They have attributed this anomaly to the reflection of the pericardium from the dilated pulmonary artery. Since then, Hudson (1965)(2) and Ikeda et al. (1969)(4) reported this anomaly. The case of Hudson was associated with a combination of pulmonary stenosis, foramen ovale and endocarditis, whereas Ikeda et al. have reported 20 cases of the tetralogy of Fallot with a pulmonary diverticulum and five cases of the tetralogy of Fallot with patent ductus arteriosus. They attributed this anomaly to a partial closure of the 6th branchial artery, resulting in partial patency of the ductus arteriosus (blind ductus). Their incidence of the pulmonary diverticulum in the tetralogy of Fallot, inclusive of patent ductus arteriosus, was 9.7% (25/259). According to our review of the angiograms of the tetralogy of Fallot, the pulmonary diverticulum was much more frequently observed than in any of the reports hitherto reported, and all the diverticula were located at the main pulmonary artery or at or near its bifurcation, where a ductus arteriosus usually arises. In our series, 34 diverticula were found either at the main pulmonary artery or at the bifurcation, the overall incidence being 30.9%. Moreover, our study has shown that pulmonary diverticulum was more frequently associated with the right aortic arch (48%) than with the left arch (25.9%). In addition to the pulmonary diverticulum, patent ductus arteriosus was also observed in 3 cases of the tetralogy of Fallot. In one of the cases of the tetralogy of Fallot with right aortic arch, a diverticulum-like protrusion of the arterial pouch was seen at the inferior aspect of the proximal left innominate artery directed toward the main pulmonary artery which also showed a diverticulum (Case 2).

Pulmonary diverticula were more frequently observed in the tetralogy of Fallot with supravalvular pulmonary artery stenosis or stenosis of both proximal pulmonary arteries.

These findings suggest that the etiology of the pulmonary diverticulum seems, as Taussig(8), Kerley et al.(7) and Ikeda et al.(4) have stated, related to the natural closure of the ductus arteriosus immediately after birth. The regression of the 6th as well as 4th branchial artery appears to play an important role in the development of the diverticulum in the case of pulmonary stenosis which causes abnormal flow patterns of the blood in the main pulmonary artery.

REFERENCES

1) Guntheroth W G : Tetralogy of Fallot, in Textbook of Heart Diesease in Infants, Children and Adolescents, edited by Moss A J and Adams F H, P 434, The Williams and Wilkins Co, Baltimore, 1968
2) Hudson R E B : Cardiovascular Pathology Vol II P. 209, Edward Arnold Publishers Ltd., 1965
3) Ikeda K, Ishida T, Taguchi Z, Todo K, Ohori I and Iwa I : Anomaly of the arch and its tributaries associated with tetralogy of Fallot. Jap J Thorac Surg 23 (1) : 41–47, 1970 (in Japanese)
4) Ikeda K, Yukawa M, Adachi H, Chikazawa M and Wada J : Pulmonary Diverticulum, Jap J Thorac Surg 22(7) : 526–529, 1969 (in Japanese)
5) Kees S and Somerville J : Aortography in Fallot's tetralogy and variants. Brit Heart J 31 : 146–153, 1969
6) Keith D J, Rowe R D and Vlad P : Heart disease in infancy and childhood, P609, 2nd ed, The MacMillan Co., New York, 1968
7) Kerley P and Strickland B : Blind ductus arteriosus. Brit J Radiol 34 : 291–294, 1961
8) Krause D A : Unusual angiographic appearance of a non-patent ductus arteriosus. Amer J Roentgenol 112(1) : 50–51, 1971
9) Rudhe U, Kjellberg S R, Mannheimer E and Jonsson B : Diagnosis of Congenital Heart Disease. 2nd ed., The Year Book Publishers, Inc Chicago, 1959
10) Satyanarayana Rao B N, Anderson R C and Edwards J E : Anatomic variations in the tetralogy of Fallot, Amer Heart J 81(3) : 361–371, 1971
11) Swischuk E L : Plain film interpretation in congenital heart disease, P. 124, Lea and Febinger, Philadelphia, 1970