Congenital Unilateral Absence of the Right Pulmonary Artery and Complex of Fallot

A Rare Association

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SUMMARY

A 16-month-old girl with a diagnosis of severe valvular pulmonary stenosis, ventricular septal defect and unilateral absence of the right pulmonary artery proven by cardiac catheterization, angiography and surgery is presented. The right lung was smaller than the left one on chest X-ray, and aortography showed that it was supplied by a rudimentary circulation arising from intercostal branches of the descending thoracic aorta.

Additional Indexing Words:

Unilateral absence of one pulmonary artery Tetralogy of Fallot

CONGENITAL unilateral absence of one pulmonary artery (PA) has been described as an isolated malformation[1] or in association with other congenital heart diseases.[2] Unilateral absence of one PA, usually the left one, has been more frequently described in association with tetralogy of Fallot (TF).[2-9]

The present case has a congenital unilateral absence of the right PA associated with valvular pulmonary stenosis and ventricular septal defect (VSD) diagnosed by cardiac catheterization and angiography.

CASE REPORT

A 16-month-old white female was admitted for cardiac surgery. She was born at term (3.840 Kg and 50 cm). Pregnancy and delivery were uncomplicated. Significant cyanosis was seen only in the first hours; after that, a mild cyanosis accentuated by exertion or crying was described. Dyspnea was noticed when she started crawling, and became more accentuated

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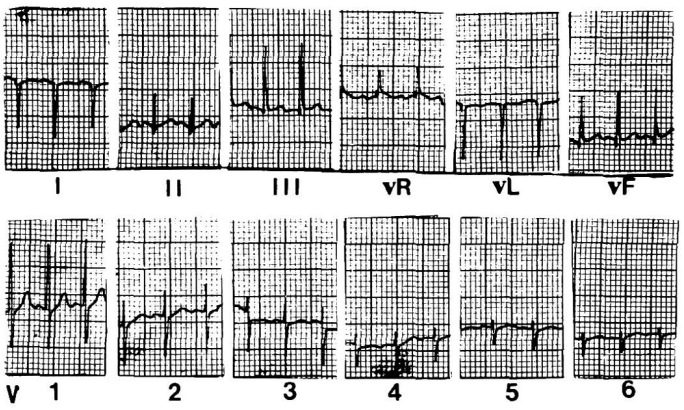


Fig. 1. Preoperative electrocardiogram. Right axis deviation (+150°) and right ventircular enlargement are observed.

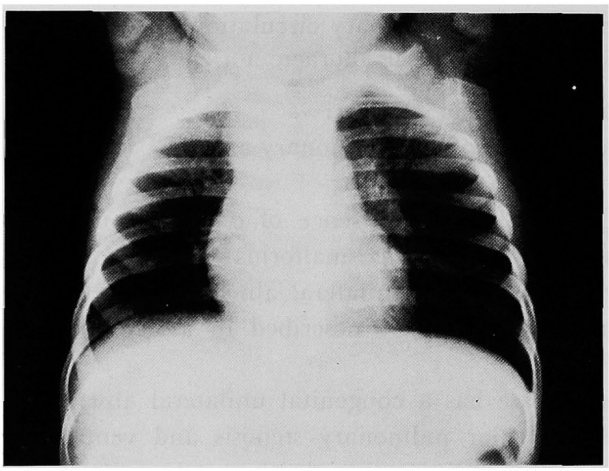


Fig. 2. Chest X-ray in the postero-anterior view. The heart is slightly enlarged due to an enlargement of the right cavities. Increased main pulmonary artery branches and strikingly diminished right lung vascular markings. Right aortic arch. Right lung smaller than the left one.

when she started walking.

Physical examination revealed: 1) a 78 cm tall girl weighing 11 Kg; 2) a mild cyanosis of the lips, fingers and toes, without clubbing; 3) the lungs were clear but the normal murmur was slightly diminished in the right lung; 4) the second heart sound was soft and not split in the pulmonic area; 5) a 5/6 rough systolic murmur was heard in the same area and was also audible along the left sternal border, aortic area and lower right sternal border. The electrocardiogram (ECG) and the chest X-ray are shown in Figs. 1 and 2. Cardiac catheterization and angiography were performed when she was

Table I. Pressure Data in mmHg

RA	LA	RV	LPA/PA	Ao	LV
(9)	(9)	100/10	22/10 (16)	100/59.5 (70)	100/10

Ao=aorta; LA=left atrium; LPA=left pulmonary artery; LV=left ventricle; PA=pulmonary artery; RA=right atrium; RV=right ventricle; ( )=mean pressure.

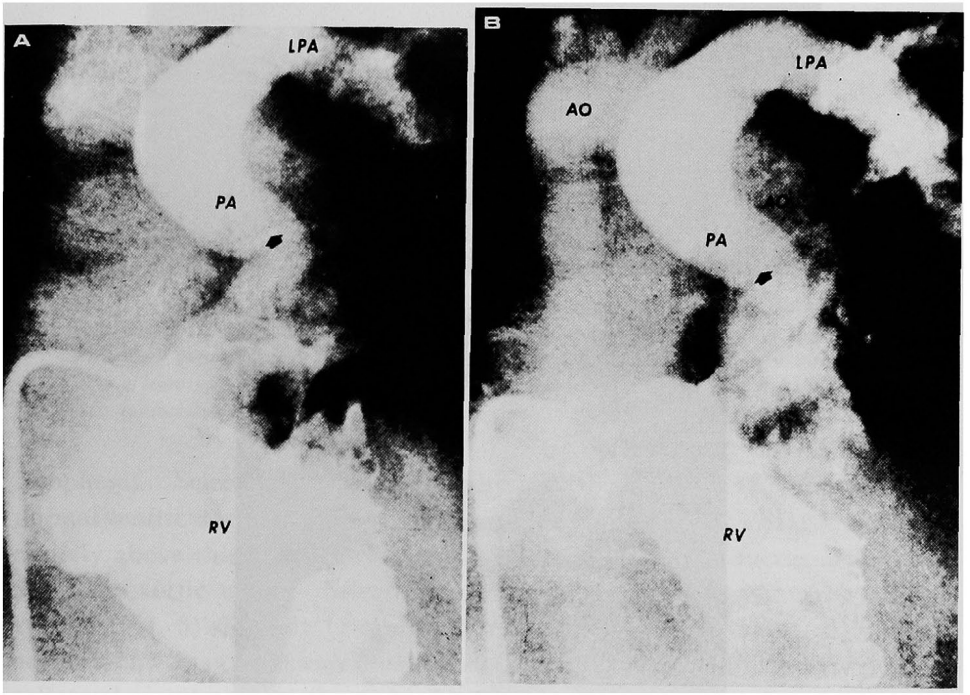
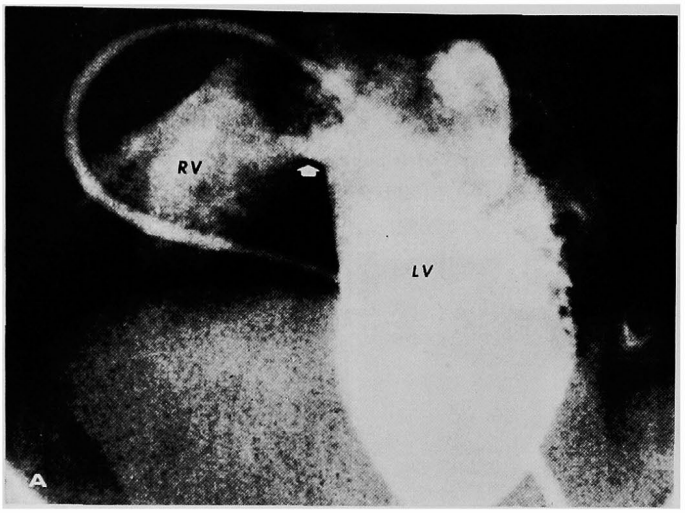


Fig. 3. Right ventriculogram in the postero-anterior craniocaudal 40° view. A: diastole, B: systole. The main pulmonary artery is in continuity with the left pulmonary artery. The right pulmonary artery is absent. The pulmonary valve is severely stenotic (indicated by arrow). The aorta is opacified through a ventricular septal defect. Ao=aorta; LPA=left pulmonary artery; PA=main pulmonary artery; RV=right ventricle.

14 months old.

Pressure data are shown in Table I. Oximetry revealed no left-to-right shunt and left ventricular (LV) oxygen saturation was 88%. Selective right ventricular (RV) angiography (Figs. 3A and 3B) revealed: 1) a hypertrophic ventricle with mild hypocontractility; 2) a severe stenosis of the pulmonary valve with a small annulus and no evidence of subpulmonary stenosis; 3) a dilated PA entirely above this ventricle leading to only the left PA; 4) a subpulmonary conus; 5) no evidence of the right PA; 6) a paramembranous VSD; 7) a small atrial septal defect (ASD) ostium secundum type (seen during



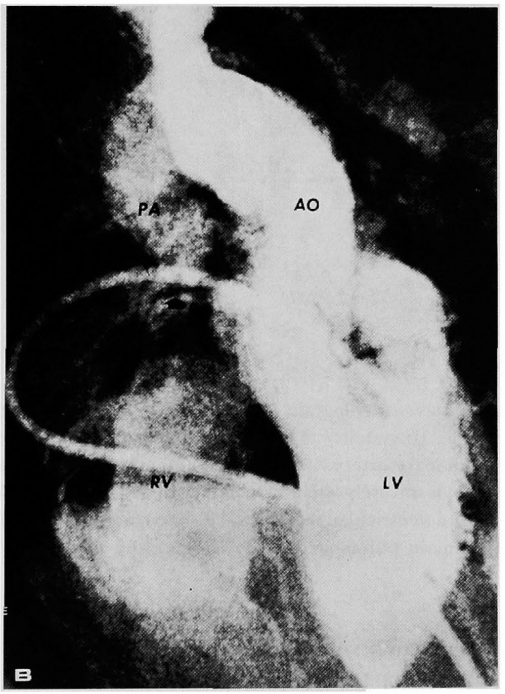


Fig. 4. Left ventriculogram in the hepato-clavicular view. A: diastole. A paramembranous ventricular septal defect is seen (indicated by white arrow) and through it the right ventricle is opacified. B: systole. The black arrow indicates the severely stenotic pulmonary valve that is at a higher level than the aortic valve. Ao=aorta; LV=left ventricle; PA=main pulmonary artery; RV=right ventricle.

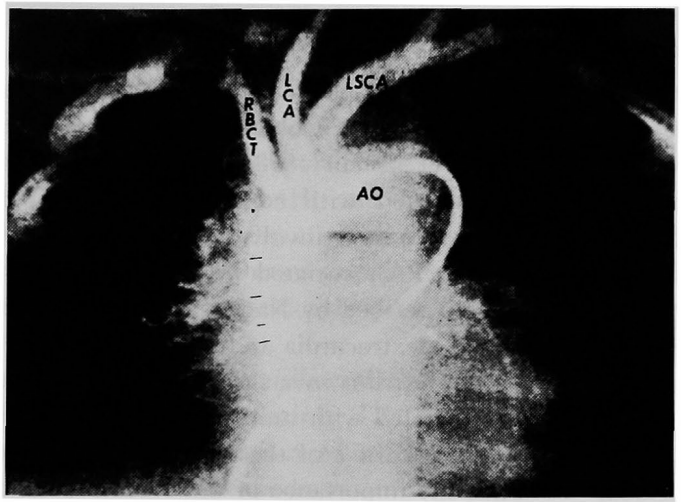


Fig. 5. Selective angiogram of the ascending aorta in the postero-anterior craniocaudal 40º view. The aortic arch is located to the right. Very poor systemic circulation from the descending aorta to the right lung is seen (indicated by small black arrows). There is no evidence of the right pulmonary artery.

levophase). Selective LV angiography (Figs. 4A and 4B) showed: 1) normal ventricular contractility; 2) a large paramembranous VSD; 3) an aorta entirely above this ventricle with no evidence of a patent ductus arteriosus; 4) no subaortic conus. Selective ascending aorta and aortic arch angiography (Fig. 5) showed: 1) a normal and tricuspid aortic valve; 2) a right aortic arch; 3) normal subclavian and carotid arteries with no evidence of collateral circulation to the right PA; 4) the presence of small systemic collateral arteries from the descending thoracic aorta to the right lung.

A right pulmonary vein wedge angiogram showed no evidence of the right PA. Surgery performed through a median sternotomy was followed by: 1) a vertical incision through the RV outflow, pulmonary valve annulus and main PA; 2) closure of the VSD with a pericardial patch; 3) closure of the ASD by direct suturing. The pulmonary valve was bicuspid and no evidence of the right PA was observed. The postoperative course was uncomplicated. The postoperative ECG showed a persistent right bundle branch block.

Cardiac catheterization 17 months afterwards showed: 1) PA pressure 25/5 mmHg; 2) a 15 mmHg gradient through the pulmonary valve; 3) RV pressure 40/5; LV pressure 100/8 mmHg. No left-to-right shunt was seen and LV saturation was 91%.

Discussion

Congenital unilateral absence of one PA as an isolated malformation has more frequently been described as involving the absence of the right PA, as demonstrated by Shakibi et al[1] in 1978. TF is the congenital heart disease most frequently associated with congenital unilateral absence of one PA. Most of the cases reported involve absence of the left PA.[2)-9)] However, absence of the right PA associated with TF has very rarely been reported.[2),3),8),10)] The case described by Nadas et al[3] in 1953 was the first one and it was associated with dextrocardia and situs inversus. It is difficult to determine if dextrocardia and situs inversus were present in other cases.

Only 4 cases of TF associated with unilateral absence of the right PA have been reported,[2),3),8),10)] and in 2 of these the anatomical details were poorly described.[2),8)] Thus the importance of this further detailed report.

Although our case cannot be considered embryologically as TF due to to the absence of infundibular pulmonary stenosis,[11)] it could be considered as being a complex of Fallot from the didactic and practical points of view. Two cases described by Nadas et al[3] (absent left PA) showed only valvular pulmonary stenosis and VSD, as in our case (absent right PA).

Another interesting fact reported by Emanuel and Pattinson[5] is the frequent association of a right aortic arch and unilateral absence of the left PA, which has not been described, to the best of our knowledge, in cases of unilateral absence of the right PA. An explanation for this would be that involution of the proximal segment of the sixth aortic arch during fetal life would lead to the absence of one PA excluding any other type of anomalous origin.[12)]

A report published by Emanuel and Pattison[5] in 1956 showed a defect in the development of the bulbus cordis giving rise to lesions of the upper part of the ventricular septum and RV infundibulum, as well as absence of the left PA in all cases of TF described up to that time. In the same report[5] the authors reviewed 19 cases of absence of one PA associated with normal development of the bulbus cordis: 8 hearts were normal and in the remaining 11 (bulbus cordis not involved), the abnormality in the majority of patients was confined to the great vessels; 17 had absence of the right PA. Embryologically, it appears that in cases of abnormal development of the bulbus cordis, the absorption of the left sixth arch is affected and in a small proportion of cases this leads to absence of the left PA.

We are unable to explain a mechanism by which TF can be associated with an absent right PA.

Surgery in cases like this should be performed early, as in cases presenting

two pulmonary arteries. Turnetto et al[10] reported on a case which was successfully operated upon. More recently, Castañeda and Norwood[13] have demonstrated that unilateral absence of one PA does not affect early or late postoperative results. There are no reports about the behavior of left PA pressure in postsurgical follow-up of cases with absent right PA. In our patient, PA pressure was 25/5 mmHg and there was a mild gradient through the pulmonary valve 17 months after surgery. Postoperative PA pressure evaluation is important in all cases of unilateral absence of one PA at early and late follow-up in order to detect the possible development of pulmonary arterial hypertension.

In conclusion, congenital unilateral absence of the right PA associated with congenital heart disease, mainly TF (or complex of Fallot) has infrequently been described. Detailed anatomical evaluation is important for a correct diagnosis and surgical correction.

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