Anomalous origin of one pulmonary artery branch from ascending aorta ("so-called hemitruncus"): report of an additional case treated surgically

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The aortic origin of one pulmonary artery branch, so-called hemitruncus, is a rare congenital anomaly with poor prognosis. In this report, an additional patient is presented. The patient, a 60-day-old male infant with the right pulmonary artery originating from the ascending aorta was operated successfully. Postoperative catheterization demonstrated normal flow to the right lung and pulmonary artery pressure decreased to normal level.

Key words: hemitruncus, anomalies-pulmonary artery, abnormalities-lung.

Origin of a pulmonary artery branch from the ascending aorta (so-called hemitruncus) was first reported by Fraentzel in an adult patient in 1868[1]. Approximately 90 patients with anomalous origin of one pulmonary artery (PA) from the ascending aorta have been described in the literature[2-9]. The malformation is more often recognized in neonates and infants as relentless heart failure. The most prominent clinical feature in patients surviving into adulthood is dyspnea on exertion and recurrent hemoptysis. Early diagnosis and corrective surgery is the treatment of choice to avoid pulmonary vascular obstructive disease (PVOD).

We report an additional case of anomalous origin of one pulmonary artery from the ascending aorta, diagnosed by cardiac catheterization and angiography, who underwent successful hemodynamic repair.

Case Report

A male infant weighing 4,100 g was born at term following an uncomplicated pregnancy. He initially seemed well, but was taken to another hospital at four weeks of age because of feeding difficulties and dyspnea. He was referred to our hospital at 55 days of age because of congestive heart failure. He was found to be in severe congestive heart failure and was treated with digitalis and diuretics. The precordium was hyperdynamic. A grade 3/6 continuous murmur was heard along the left sternal border. The liver's edge was 4 cm below the right costal margin. The arterial PO_2 was 64 mmHg with an oxygen saturation of 72%. Despite medical therapy, the patient's heart failure did not improve.

Two-dimensional echocardiography revealed the origin of the right pulmonary artery from the ascending aorta, patent ductus arteriosus (PDA) and moderate tricuspid regurgitation. Cardiac catheterization (Table I) and angiography (Fig. 1) confirmed the diagnosis. The patient was then transferred to the Cardiac Surgery Department, and the operation was undertaken on the following day. At operation, via median sternotomy, it was observed that the right PA originated from the posterior wall of the ascending aorta. The PDA was exposed and occluded with a snare. By means of standard bicaval cannulation and hypothermic (24 °C) cardiopulmonary bypass, main PA, right PA and ascending aorta were mobilized. First, the aorta was opened transversely on its anterior surface just at the level of the right PA connection. The right PA orifice was found to be slightly stenotic and was excised with a button, containing a part of the posterior aortic wall. The right PA orifice

was enlarged with an anterior autologous pericardial patch to ensure construction of a widely patent tension-free anastomosis. It was then sutured to the main PA, placed posteriorly to the aorta. The aortotomy was closed primarily, and the PDA was divided and oversewn (Fig. 2).

The patient tolerated the operation well. He was discharged on the 9th postoperative day receiving digoxin only. At cardiac catheterization six months postoperatively, the PA pressure was normal and there was no gradient across the anastomosis (Figs. 3, 4). On follow-up 24 months after surgery, he appeared to be a normal, very active boy.

Table I. Preoperative and Postoperative Catheterization Data

	Pre-op	Post-op (13th month)
Pressure (mmHg)		
MPA	76/36 (51)	27/4 (13)
RPA	66/33 (50)	26/4 (13)
RV	87	27
LV	91	97
Ao	91/64 (74)	97/60 (68)
Oxygen saturation		
MPA	53	62
RPA	83	84
RV	44	–
LV	90	97.1
Ao	75	98

MPA: Main pulmonary artery. RPA: Right pulmonary artery. RV: Right ventricle. LV: Left ventricle. Ao: aorta. The value in parentheses indicates mean pressure.

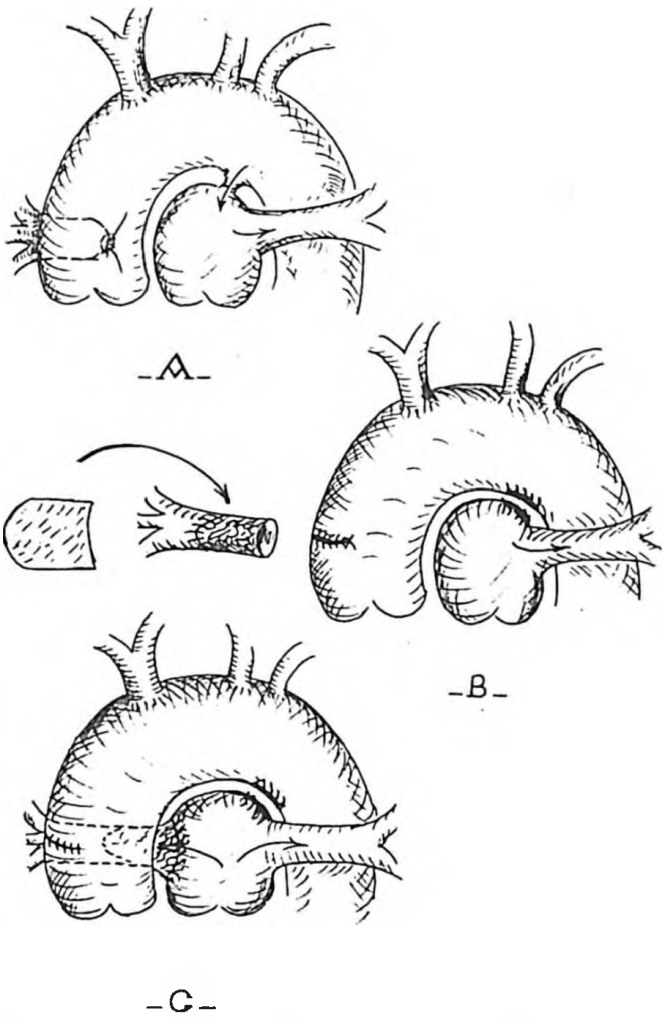
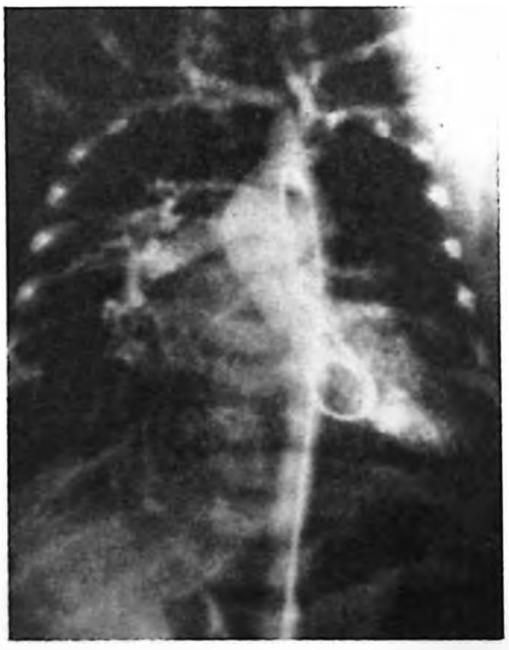
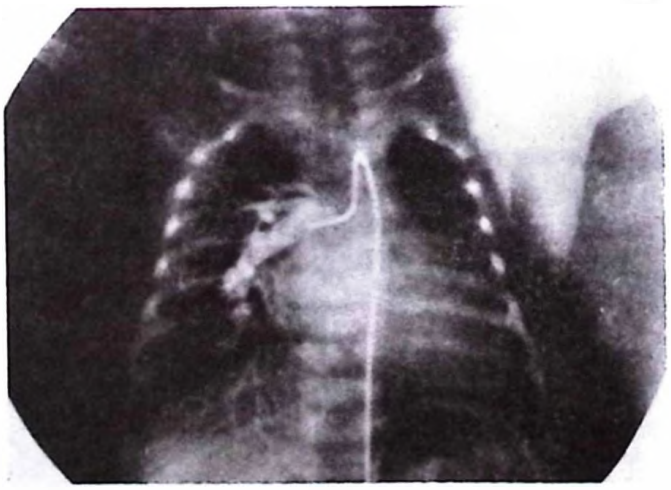


Fig. 2. Operation technique. A: preoperative view. B: the anomalous right PA is detached from the aorta with a portion of aortic wall, and the right PA orifice was enlarged with an autologous pericardial patch. C: the right PA is sutured to the main PA behind the ascending aorta.



(a)



(b)

Fig.1. Preoperative angiogram (A and B) showing anomalous origin of right pulmonary artery (RPA) branch from the ascending aorta.

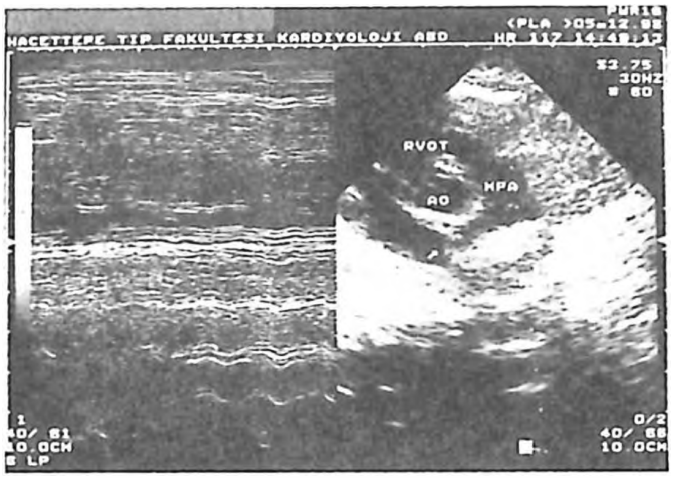


Fig.3. Parasternal short-axis view showing the right ventricular outflow tract (RVOT) and the pulmonary artery branches after correction (MPA: main pulmonary artery, LPA: left pulmonary artery, RPA: right pulmonary artery, Ao: aorta).

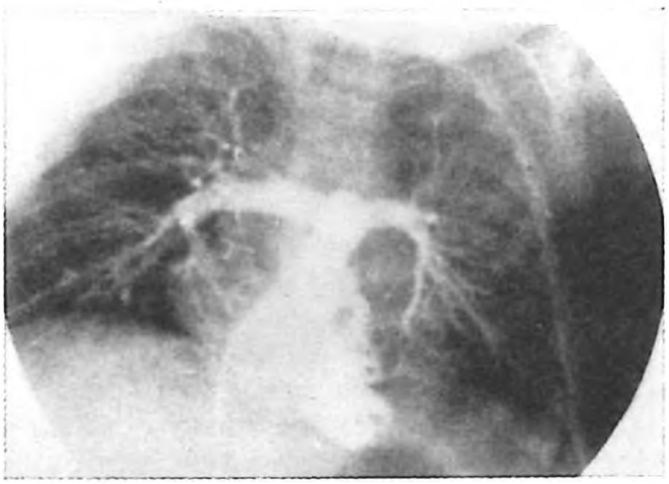


Fig. 4. Postoperative right ventricular control angiogram showing uniform distribution of pulmonary blood flow.

Discussion

Anomalous origin of one pulmonary artery branch from the ascending aorta, also called hemitruncus arteriosus, is a rare, congenital cardiovascular malformation with poor prognosis if it is not treated surgically in early infancy[3,4,7]. Although it was first described in an adult patient by Fraentzel[1], the anomaly is more often recognized in neonates and infants; only eight patients have been diagnosed during adulthood[6,8,9]. The most common associated anomaly when the right PA is involved is a PDA, which always occurs contralaterally to the anomalous pulmonary artery. In the case of the left PA branch originating from the ascending aorta, tetralogy of Fallot is the most frequently associated malformation[4].

Penkoske and coworkers[4] stated that analysis of the literature on origin of a pulmonary artery branch being from the ascending aorta is difficult, because at least three different cardiovascular malformations have been classified as such name:[1] origin of the right or left PA from the ascending aorta;[2] absence of the proximal portion of either pulmonary artery, with the distal pulmonary artery branch being supplied via PDA;[3] absence of a proximal pulmonary artery branch, the distal pulmonary artery branch being supplied via an aorticopulmonary collateral artery, not via a PDA.

Although diagnosis of the anomaly has usually been made at cardiac catheterization[2-7], echocardiography has also been useful in infants[5,9]. Recently it was reported that the diagnosis could be made by computed tomography[6] or by magnetic resonance imaging[8].

To date, different kinds of procedures have been performed for correction. The first corrective procedure was published by Caro and coworkers in 1957[2]. An Ivalon graft was interposed between the right PA and the main pulmonary artery (MPA) in that case. Unfortunately the patient died after completion of the operation. The first successful operation using the same technique but with a Dacron graft was reported by Armer and coworkers in 1961[10]. In 1967; Kirkpatrick and coworkers[11] reported the first case in which primary anastomosis was performed between the aberrant right pulmonary artery and the MPA. Since than, many patients have undergone surgical correction in relation to their cardiac anomalies in a one-or two-stage fashion with or without the aid of cardiopulmonary bypass and hypothermia.

Because the patients surviving into adulthood are at risk for developing pulmonary vascular obstructive disease, surgical repair should be performed as early as possible in order to prevent death from congestive heart failure or the development of irreversible PVOD[1,2,8,9]. In patients with an anomalous origin of one pulmonary artery branch from the ascending aorta, the pulmonary hypertension in the main pulmonary artery and the contralateral pulmonary artery branch seems to be reversible when correction is performed in infancy[4]. However, in older children and adults, who survive to the operation, pulmonary hypertension may still persist[4,12]. Although the exact mechanism of the persistent pulmonary

hypertension in the MPA and in the protected (contralateral) lung has not been fully explained, a neurogenic crossover from the lung perfused by the aorta to the protected lung has been postulated by Griffiths and coworkers[13].

In our patient, the MPA pressure decreased from 76/36 mmHg to 27/4 mmHg after the correction. In patient with hemitruncus, diagnosis and surgical correction should be performed as early as possible to prevent death from heart failure or the development of irreversible PVOD.

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