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Coronary artery changes 3 years after reimplantation of an anomalous right coronary artery

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In this paper we report the sequelae of a patient with an anomalous right coronary artery (RCA) originating from the pulmonary artery (PA) in association with a normal heart, operated upon at the age of 13 years. Three years after the end-to-side reimplantation of the RCA, with a rim of the PA, into the aorta, the surgical result has been evaluated by cineangiography. Before operation both coronary arteries were tortuous and increased in size. Afterwards the left coronary artery showed a normalized calibre, although the RCA remained tortuous with no decrease of the internal diameter. The notable postoperative changes in shape and size of the LCA may be due to the disappearance of the steal phenomenon. The lack of involutive changes in the RCA could be explained by its thinner wall. Left ventricular wall motion, evaluated under resting conditions and during an atrial pacing stress test, was found to be normal.

Introduction

Of the four types of anomalous origin of coronary arteries, described by Soloff, the anomalous origin of the right coronary artery from the pulmonary artery is the second most common form[1]. Up till now, 25 cases have been reported. A congenital cardiac malformation associated with an anomalous right coronary artery arising from the pulmonary artery has been described in 7 cases: 3 with Fallot's Tetralogy[2–4], 3 with aortopulmonary window[5–7] and 1 with atrial septal defect[8]. Also it has been described in association with otherwise normal hearts in 18 instances[9–24]. In this paper we report one additional case with an otherwise normal heart, operated upon at the age of 13 years and reevaluated angiographically three years after corrective surgery.

Case report

A healthy 7 year-old boy was referred to our institution for further evaluation of a heart murmur, first noticed at the age of 5 years.

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On auscultation the first sound was loud and single, there was a grade 1–2/6 continuous murmur in the second and third left intercostal spaces. The second sound was variable and physiologically split. Liver and spleen were of normal size. Arterial pulses were normal. The chest X-ray revealed a slightly enlarged heart associated with a somewhat increased pulmonary circulation as well as a left-sided descending aorta. The electrocardiogram showed a sinus rhythm and no signs of hypertrophy. At cardiac catheterization, pressures in both ventricles were normal. Oxygen saturation in the right ventricular outflow tract was 81%, while that in the pulmonary artery trunk (PA) 85%. The ratio of pulmonary versus systemic flow was 1·5 : 1·0 (8·7 : 5·8 l min^{-1} m^{-2}). A cineangiogram performed after a supravalvular aortic injection revealed a dilated and tortuous left coronary artery (LCA) arising from the left aortic cusp, giving collaterals to the right coronary artery (RCA), which was tortuous and dilated throughout its course and emptied into the pulmonary artery (Fig. 1). The coronary sinus was not visualized during the venous phase of the coronary angiogram.

Although the boy remained asymptomatic a diagnostic catheterization was advised at the age of 13 years since the continuous murmur was altered into a midsystolic and middiastolic and reduced

in intensity. The oxygen saturation recorded from the PA to the right ventricle again showed a step down of 4% and a selective left coronary cineangiogram confirmed the findings of the previous angiography. Shortly afterwards he was operated under hypothermia with extracorporeal circulation. On visual inspection, the RCA was found to be dilated, vein-like and tortuous, whilst the LCA was also dilated and tortuous, but resembled an artery. The origin of the RCA was excised with a rim of the PA, an end-to-side anastomosis with the ascending aorta was performed and the PA was sutured. The post-operative period was uneventful.

At the age of 16 he had normal heart sounds and no residual murmur. The chest X-ray showed a normal heart as well as a normal pulmonary circulation. Work capacity was evaluated with a graded bicycle test with computer processing of the ECG during and after exercise[25]. The maximal workload achieved was 260 Watts. The electrocardiogram at rest, during exercise and recovery were normal.

At catheterization the pressures in RV, LV and aorta were found to be normal. The LV function and LV wall motion were investigated under resting conditions and during an atrial pacing stress test and were found to be normal as well (Table 1). Cineangiographically, the LCA appeared normal, was no longer tortuous and showed no collaterals (Figure 2). The RCA appeared less tortuous but still remained large (Figure 3). The quantitative measurements of the internal diameters of the RCA and LCA, determined by the computer based

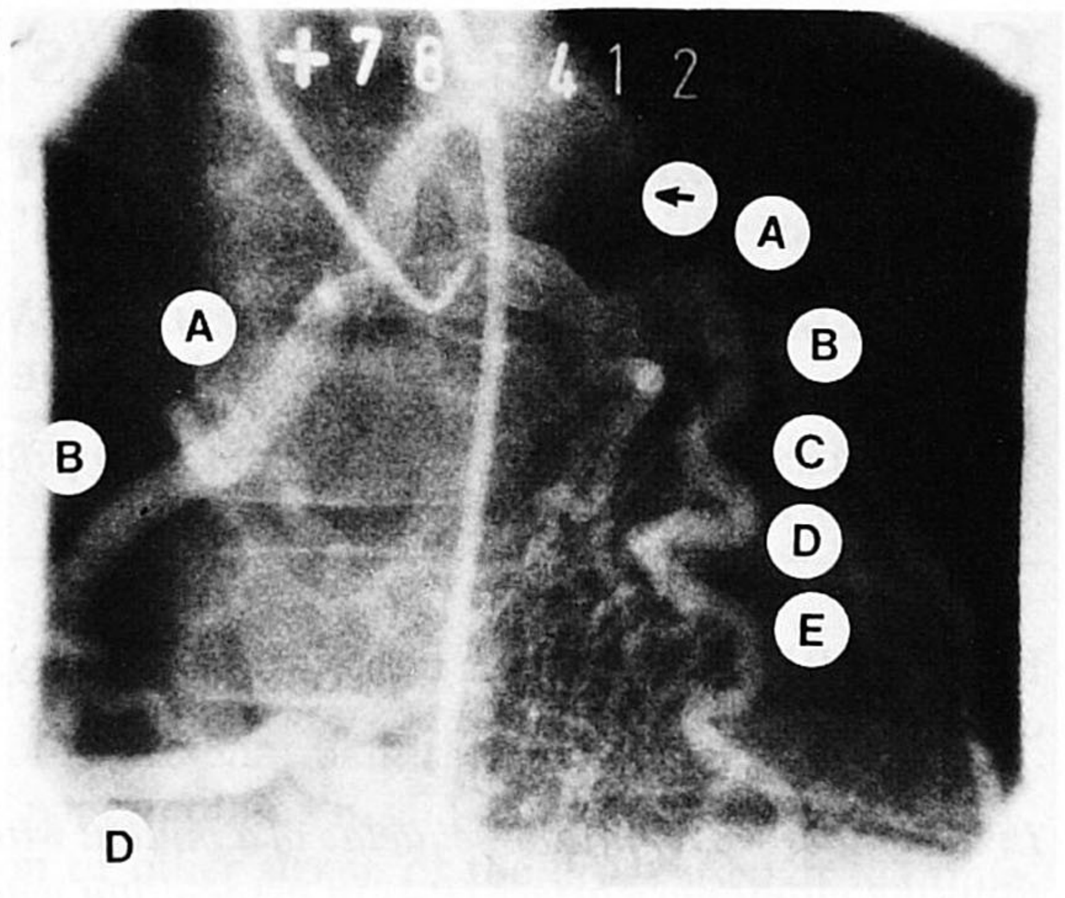
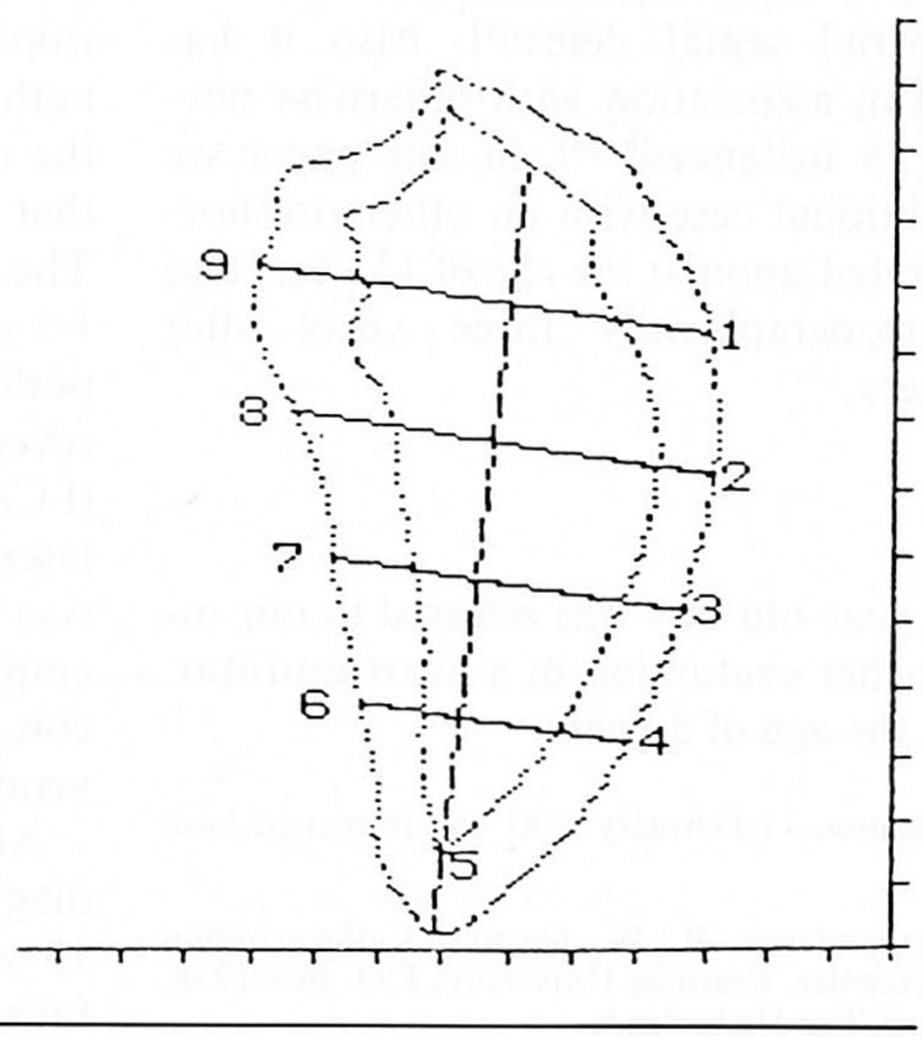


Figure 1 Anomalous RCA from the pulmonary artery (PA). RCA and LCA are both dilated and tortuous. The RCA is filled retrogradely through collaterals from the LCA and empties in the PA. The arrow indicates the PA valves. Preoperative internal diameter (m ± sd) of RCA and LCA measured by computer based angiographic analysis system: *RCA segment* A: 8·9 mm ± 0·4; B 8·2 mm ± 0·2; D: 6·9 mm ± 0·2. *LCA segment* A: 6·7 mm ± 0·3; B: 6·7 mm ± 0·1; C: 6·1 mm ± 0·1; D: 5·5 mm ± 0·4; E: 4·6 mm ± 0·4.

Table 1 Left ventricular wall motion during rest (R) and during atrial pacing (A). Values of segmental shortening were measured along 9 hemaxial cords perpendicular to the long axis of the heart. Values R were obtained at rest and values A during atrial pacing. The normal values of the different segments are given in table 'norm' (L = low; H = high). It is shown that during atrial pacing the segmental motion increases in the hypokinetic parts and normalizes in the hyperkinetic parts

Cord	R (%)	A (%)	Norm (%)
1	46	52	41–65
2	25 L	28 L	41–69
3	27 L	30 L	38–66
4	40 L	40 L	43–71
5	38	36	23–61
6	62	59	44–66
7	56 H	50 H	33–49
8	47 H	40	28–41
9	39 H	17	8–34



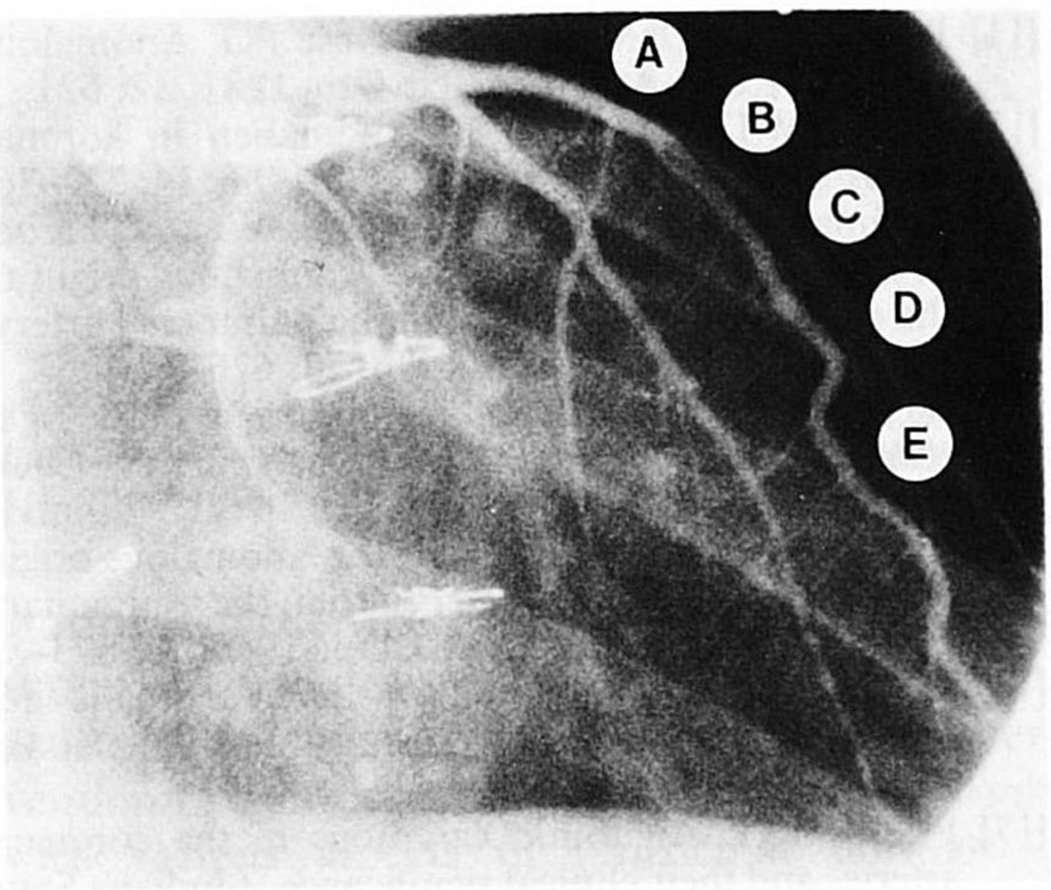


Figure 2 The shape and size of the LCA, three years following operation. Postoperative internal diameters (m ± sd) of the LCA determined by computer based angiographic analysis system: *LCA segment:* A: 3·1 mm ± 0·2; B: 2·7 mm ± 0·1; C: 2·7 mm ± 0·4; D: 2·6 mm ± 0·1; E: 2·7 mm ± 0·1.

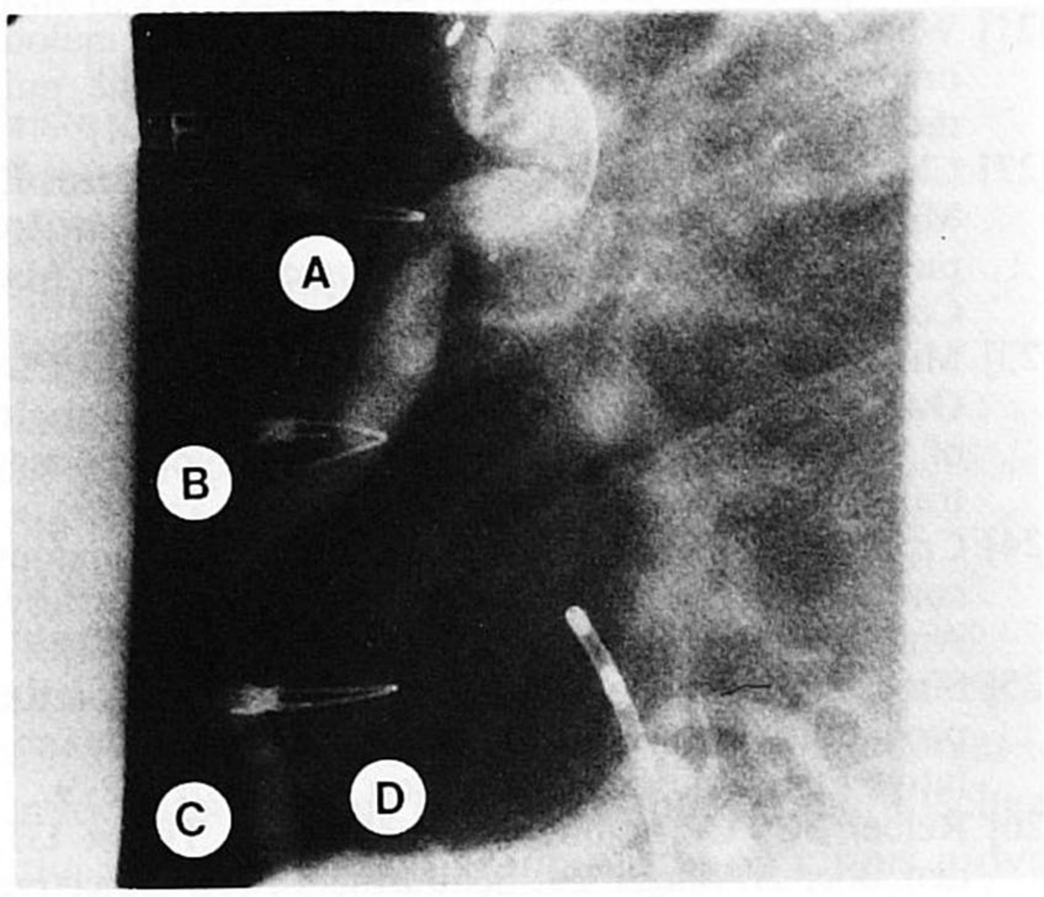


Figure 3 The shape and size of the RCA, three years following operation. Internal diameters (m ± sd) of the RCA determined by computer based angiographic analysis system: *RCA segment:* A: 12·8 mm ± 1·5; B: 7·6 mm ± 0·7; C: 8·2 mm ± 0·6; D: 6·9 mm ± 0·3.

angiographic analysis system[26], before and after surgical correction showed that while the RCA diameter of 8 mm did not decrease, the LCA had diminished from 6 to 2·7 mm in internal diameter (Figs 1–3). The coronary sinus was now normally visualized during the venous phase of the coronary angiogram.

Discussion

In contrast to the anomalous origin of the LCA, the anomalous origin of the RCA from the pulmonary artery rarely causes cardiac symptoms or electrocardiographic features of ischaemia[2,9,21–24]. However, sudden death has been reported[9,13,21,27] and when the lesion is recognized, operation is recommended[9,23,27].

In the past, four different types of surgical correction of this abnormality have been performed: (1) ligation of the anomalous RCA creating a single coronary artery system with a potential risk of death[8,12,21]; (2) ligation of the RCA with implantation of a venous graft from the aorta to the RCA[14]; (3) side-to-side anastomosis of the RCA and the aorta, with ligation of the pulmonary artery origin of the RCA[22]; and (4) reimplantation of the RCA with a narrow rim of tissue from the pulmonary artery sutured end-to-side into the aortic wall[7,9–11,13,20,23], with one late death[5]. In our patient the latter procedure was accomplished without complications and RV- and LV-function as well as LV wall motion were found to be normal (Table 1). There were remarkable differences in the appearance of the coronary arteries. The preoperative selective LCA-angiogram showed a tortuous LCA with an internal diameter of about 6·0 mm (Fig. 1), whereas after surgical correction the LCA was virtually normal in shape, showing an internal diameter of about 2·7 mm (Fig. 2). The RCA, however, remained tortuous with no reduction of the internal diameter, although collaterals between the LCA and RCA no longer were apparent and the coronary sinus was normally visualized (Fig. 3).

Since the LCA fills the RCA in a retrograde direction by way of the collaterals[27,28], given the pressure gradient a left-to-right coronary artery steal phenomenon is created which can lead to ischaemia, as shown in two other cases[22,23]. In our patient, the poor visualization of the coronary sinus by the contrast in the venous phase of the cineangiogram may corroborate this interpretation. Furthermore the increased bloodflow through both coronary arteries leads to their larger size and tortuosity. At surgery the RCA indeed looks like a vein, although postmortem histological study of the wall of the RCA when available has shown that the media while thin still resembles that of an artery[15,24]. After surgical correction, the intraluminal pressure in the RCA returns to systemic levels and the left-to-right coronary steal phenomenon is abolished, as a consequence of which the coronary sinus is normally visualized during the

venous phase of the cineangiogram. The reduced blood flow in the coronary arteries has allowed the LCA to return to normal dimensions. The lack of involutional changes in the right coronary artery could be caused by its thin wall with a structurally changed and thinned media[15,24].

In conclusion it can be said that the reimplantation of the anomalous RCA with a rim of the pulmonary artery into the aorta in this patient was successful. The LV function as examined with the bicycle exercise test and cineangiographically at rest and during atrial pacing stress test was found to be normal, while the LCA returned to normal shape.

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