Clin. Cardiol. 21, 773–776 (1998)

Anomalous Origin of the Right Coronary Artery from the Main Pulmonary Artery: Diagnosis and Management

VAHID HEKMAT, M.D., SUDHA M. RAO, M.D., MANOJ CHHABRA, M.D., MARIO CHIAVARELLI, M.D., PH.D., JOHN E. ANDERSON, M.D., DOV B. NUDEL, M.D.

Department of Pediatrics, SUNY/HSCB, Brooklyn, New York, USA

Summary: Anomalous origin of the right coronary artery (ARCA) from the main pulmonary artery (MPA) is a rare congenital anomaly, with only 18 reported cases in the pediatric age group. More than half of these had associated cardiac anomalies that masked the presence of ARCA. Conversely, in many patients with ARCA as an isolated anomaly, the diagnosis has been missed during lifetime. The only patient with an isolated ARCA who was diagnosed in infancy presented with congestive heart failure. Asymptomatic infants with ARCA from the MPA have not been previously reported. Three additional cases, two infants and a child with ARCA from the MPA, are reported in this paper. The diagnostic dilemmas and the prognosis are discussed and management is recommended.

Key words: anomalous, right coronary artery, diagnosis, management

Introduction

Anomalous origin of the right coronary artery (ARCA) from the main pulmonary artery (MPA) is a rare congenital anomaly first reported by Brooks [1] as an incidental autopsy finding. Overall, 52 such cases have been reported in the literature,[1–8] but only 18 were reported in the pediatric age group (< 18 years). In this group, 10 patients had associated cardiac defects, including aortopulmonary window in 5; atrial septal defect in 2; tetralogy of Fallot in 2; and ventricular septal defect, patent ductus arteriosus, and pulmonic stenosis each in 1 patient. Of the eight patients in whom ARCA was an isolated anomaly, one presented with congestive heart failure, four were asymptomatic but had heart murmurs considered significant, and three were diagnosed at autopsy. Four of the patients were diagnosed in infancy and three of these had additional cardiac anomalies.[9–11] The only patient with an isolated ARCA who was diagnosed in infancy presented with congestive heart failure.[12] No patients with ARCA from the MPA who were asymptomatic were previously diagnosed in infancy. In this paper, three additional cases, two infants and one child with isolated ARCA from the MPA, are reported. All three were asymptomatic and were diagnosed by two-dimensional echocardiography and Doppler studies. Their management is discussed and a review of the literature is presented.

Address for reprints:

Dov B. Nudel, M.D.
SUNY/HSCB Box 1200
Department of Pediatrics
450 Clarkson Avenue,
Brooklyn, NY 11203-2098, USA

Received: February 24, 1998
Accepted with revision: May 11, 1998

Case No. 1

A 9-month-old African American girl was referred for evaluation of a heart murmur. Birth and subsequent history were unremarkable, and she has been asymptomatic regarding her cardiovascular system.

On physical examination, weight was 7.5 kg (5th percentile), height was 71 cm (25th percentile), heart sounds were normal, and there was a grade II–III/VI continuous murmur at the left upper sternal border. The rest of the cardiovascular examination was normal. The electrocardiogram (ECG) showed a QRS axis of 80°, deep Q waves of 4–8 mm in leads II, III, and aVF, but the width of the Q waves was only 1 mm. There was mild left ventricular hypertrophy by voltage criteria. The two-dimensional (2-D) echocardiogram showed a right coronary artery (RCA) in direct communication with the MPA (Fig. 1). The RCA was uniformly dilated and tortuous, and Doppler study showed diastolic reversal in the descending abdominal aorta. Color Doppler showed retrograde flow from the RCA into the MPA (Fig. 2). Cardiac catheterization and angiography demonstrated anomalous origin of the RCA from the MPA (Fig. 3) with a significant oxygen step-up in

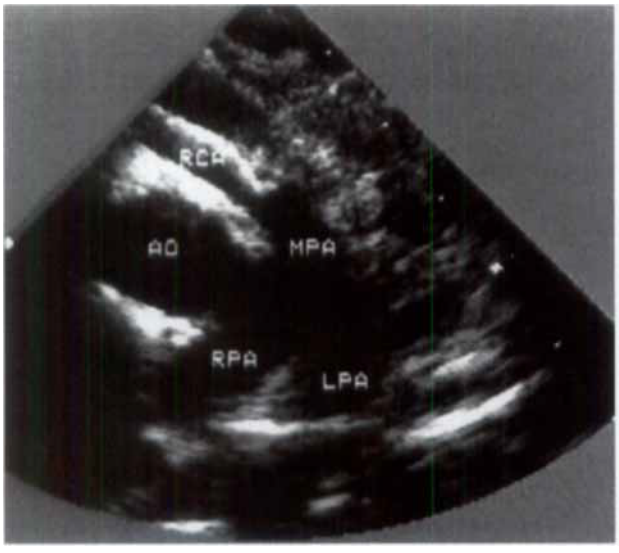

FIG. 1 Two-dimensional echocardiographic study, parasternal short axis view: Note the direct communication of the RCA with the MPA. Ao = aorta, RCA = right coronary artery, MPA = main pulmonary artery, RPA = right pulmonary artery, LPA = left pulmonary artery.

FIG. 2 Color Doppler study of the same view as in Figure 1. Legend as in Figure 1. Note retrograde flow from RCA into MPA.

the MPA compatible with a Qp/Qs ratio of 1.9 to 1.0. Pulmonary artery pressure was normal. The infant was submitted for surgery during which the RCA was transsected from the MPA and reimplanted into the ascending aorta. A select angiogram of the RCA 1 year after surgery showed unobstructed flow in the transplanted coronary artery (Fig. 4). Follow-up period was 5 years and 10 months, and the patient has remained asymptomatic.

Case No. 2

A 7-month-old African American boy with a history of obstructive sleep apnea was referred for an echocardiographic Doppler study to evaluate the presence of pulmonary artery hypertension. Prenatal history indicated maternal diabetes mellitus and hypertension. The infant was born prematurely at a gestational age of 36 weeks because of maternal preeclampsia.

Past medical history was otherwise unremarkable, and the infant was asymptomatic regarding the cardiovascular system.

Physical examination was unremarkable: weight was 11 kg, height was 80 cm, both at the 25–50th percentile. Cardiovascular examination showed a faint grade I/VI systolic ejection murmur at the left upper sternal border. The ECG and the chest x-ray were normal.

The 2-D echocardiogram, Doppler study, and angiography showed pictures similar to those of Patient No. 1, but the RCA was less dilated and the left-to-right shunt was smaller.

Surgery was performed similar to that in Patient No. 1. The postoperative course was unremarkable and during a follow-up period of 4 years and 5 months the patient has remained asymptomatic.

Case No. 3

A 5-year-old African American girl was referred for evaluation of a heart murmur. She had been asymptomatic regarding her cardiovascular system.

On physical examination, weight and height were both at the 90th percentile. There was a grade II/VI soft systolic ejec-

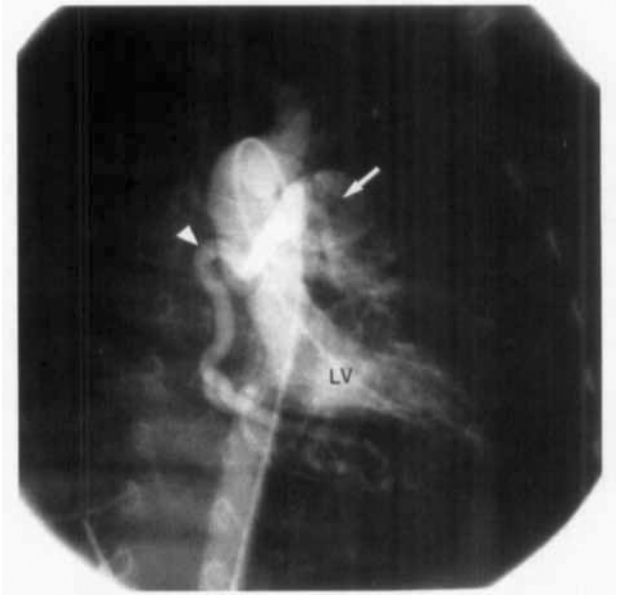

FIG. 3 Left ventricular angiogram. Contrast has mostly cleared from left coronary artery. Note large and tortuous RCA seen entering MPA. Arrow indicates MPA. Arrow head shows RCA. LV = left ventricle. Other abbreviations as in Figure 1.

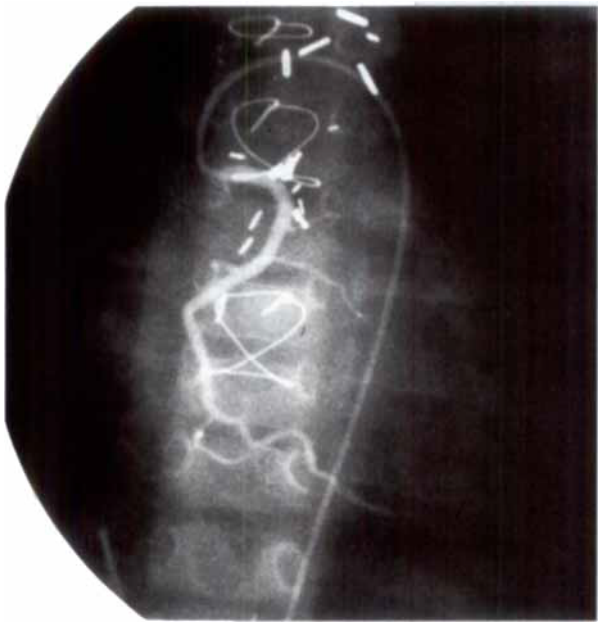

FIG. 4 Selective right coronary artery (RCA) angiogram. Note the origin of the reimplanted RCA into the ascending aorta. The RCA is less dilated and less tortuous 1 year after reimplantation.

tion murmur best heard at the left upper sternal border. The rest of the cardiovascular examination was normal.

The ECG and the chest x-ray were normal. The 2-D echocardiogram and the angiogram were similar to those of the previous two patients. There was a small oxygen step-up in the MPA compatible with a Qp/Qs ratio of 1.3 to 1.0, and pulmonary artery pressures were normal. Cardiac surgery was performed similar to the previously described patients. The postoperative course was unremarkable. Follow-up period was 2 years and 8 months, and the patient has been asymptomatic.

Discussion

Many congenital cardiac anomalies are associated with abnormalities of coronary artery origin. Most of these, however, are of abnormal origin of a coronary artery from the aorta.[13] Anomalous origin of the RCA from the MPA is a rare congenital anomaly, but since many patients reported in the literature, as well as the three patients reported in this paper, are asymptomatic and their murmurs are inconspicuous, the true incidence of this anomaly is not known. The fact that all three patients were diagnosed within a 3-year period in a modest size program such as ours may be a coincidence, but may also indicate a higher incidence than that previously reported.

Among the 18 pediatric patients, the incidence of associated cardiac anomalies was 67% and included a wide spectrum of anomalies. However, 5 of the 12 associated anomalies were aortopulmonary windows. It is difficult to explain a common embryologic defect for both ARCA and aortopulmonary window. A defect in truncal septation has been previously proposed, but is unlikely as truncal septation occurs before the appearance of coronary orifices.[14] Another theory for this association is the development of accessory coronary orifices, of which two become prominent. However, Bogers *et al.*[15] have shown that the setting of the coronary orifices is determined by the epicardial coronary network and that the coronaries grow into rather than out of the aorta.

The diagnosis of ARCA from the MPA in all pediatric and adult patients was established during cardiac catheterization performed primarily for the associated lesions, or intraoperatively as part of the complete surgical repair. Most patients with isolated ARCA from the MPA were also diagnosed by cardiac catheterization performed to evaluate either a continuous heart murmur with or without cardiomegaly[3, 8, 16–19] or unexplained congestive heart failure.[5, 12, 17, 20] In the adult patients, angiography was most commonly performed because of angina and suspected atheromatous coronary artery disease.[5, 8, 16, 18] Although the diagnosis of the three patients in this report was established by 2-D echocardiography and Doppler studies, we performed a cardiac catheterization and angiography in order to visualize the entire length and the extent of tortuosity of the RCA, assess the degree of shunting, and measure pulmonary artery pressure. It is quite likely that with increasing experience with this anomaly, the diagnosis by echocardiography and Doppler studies may suffice for a complete anatomic evaluation and surgery.

The three patients described in this paper highlight a considerable diagnostic dilemma as they were all asymptomatic, and the murmurs, at least in Patients No. 2 and 3, were inconspicuous and could have passed as a functional heart murmur of the pulmonary ejection type. The question can be raised whether all patients with such a murmur should have echocardiographic and Doppler studies to identify the few with ARCA from the MPA. This may not be cost effective, but it is important to identify these patients and to intervene surgically. Patient #1 in this report had a significant left-to-right shunt, and the indications for surgery are clear. One might argue about the indications as well as the timing of surgery in the other two patients who did not have large shunts and were asymptomatic. We recommended surgery for several reasons. First, with time and the patient's growth, the coronary arteries may continue to dilate and eventually increase in size, resulting in a larger shunt. Second, although some patients are asymptomatic, others may develop myocardial ischemia and sudden death. Wald *et al.*[18] reported a 17-year-old boy with this anomaly and ECG changes of myocardial ischemia who eventually died suddenly. Lerberg *et al.*[21] described a 2-year-old boy who was found dead in his crib, as well as two additional patients: an 11-year-old girl and a 72-year-old man who sustained a cardiac arrest. In all three cases, ARCA from the MPA was the only cardiovascular abnormality found at autopsy. Autopsy findings from other patients with ARCA who died suddenly showed no significant abnormal changes in the right ventricle, but left ventricular myocardium showed widespread pathologic findings including fibroelastosis,[18, 22] ischemia,[14, 18, 21] and infarction.[5, 18] The underlying pathophysiology for these changes is considered to be the left-to-right

shunt, which causes a steal from the left coronary artery into the pulmonary artery due to the low resistance in the latter. Finally, it is reasonable to assume that with advancing age and the development of atherosclerosis the patients will benefit more from a two-coronary artery system than from one. Therefore, we recommended transplanting the ARCA from the MPA to the ascending aorta at the time the diagnosis is made, as previously recommended by Tingelstad *et al.*[23] This will prevent the steal from the RCA with its deleterious consequences and establish a two-coronary artery system. Since significant experiences with coronary artery surgery in infants have been accumulated following the arterial switch operation for transposition of the great arteries, this surgical procedure carries a low risk and we recommend early surgical intervention even when patients are asymptomatic. The reimplanted RCA (Fig. 4) is considerably smaller and less tortuous compared with the same ARCA before surgery (Fig. 3). Although this artery is patent 1 year after surgery, long-term follow-up will determine the incidence of possible stenosis in the reimplanted vessel.

References

1. Brooks H: Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: With some remarks upon the effects of this anomaly in producing cirsoid dilatation of the vessels. *J Anat* 1885;20:26–29
2. Roberts WC: Major anomalies of coronary arterial origin seen in adulthood. *Am Heart J* 1986;111:941–963
3. Moss RL, Backer CL, Zales VR, Florentine MS, Mavroudis C: Tetralogy of Fallot with anomalous origin of the right coronary artery. *Ann Thorac Surg* 1995;59(1):229–231
4. Nakano M, Emoto H, Koyanagi K, Okuyama H, Saito F, Kurosawa H: Report of a case of the anomalous origin of the right coronary artery from the pulmonary artery with atrial fibrillation and bradycardia. *J Jpn Assoc Thorac Surg* 1993;41(3):479–485
5. Shah RM, Nanda NC, Hsiung MC, Moos S, Roitman D: Identification of anomalous origin of the right coronary artery from the pulmonary trunk by color flow mapping. *Am J Cardiol* 1986;57:366–367
6. Tosovsky J, Rohac J: Anomalous origin of the right coronary artery from the pulmonary artery. *Rozhledy V Chirurgii* 1993;72(5):199–200
7. Tuma S, Hucin B, Reich O, Voriskova M, Radvansky J: Anomalous origin of the right coronary artery from the pulmonary artery. Preoperative diagnostic finding in the aortopulmonary window. *Ceskosloenska Pediatrie* 1990;45(9):543–545
8. Ueeda M, Yamada N, Shimizu A, Hina K, Terasaka R, Saito D, Tsujii T, Yoshida H, Haraoka S: A case of anomalous origin of the right coronary artery from the pulmonary trunk. Imaging of abnormal flow by Doppler color flow mapping. *J Cardiol* 1998;18:583–587
9. Brouwer MH, Beaufort-Krol GC, Talsma MD: Aortopulmonary window associated with an anomalous origin of the right coronary artery. *Int J Cardiol* 1990; 28(3):384–386
10. Donaldson RM, Raphael MJ, Radley-Smith R, Yacoub MH: Angiographic diagnosis of anomalous origin of the right coronary artery from the pulmonary artery. *Br J Radiol* 1983;56:17–19
11. Luisi SV, Ashraf MH, Gula G, Radley-Smith R, Yacoub M: Anomalous origin of the right coronary artery with aortopulmonary window: Functional and surgical considerations. *Thorax* 1980;35(6):446–448
12. Vairo U, Marino B, De Simone G, Marcelletti C: Early congestive heart failure due to origin of the right coronary artery from the pulmonary artery. *Chest* 1992;102(5):1610–1612
13. McManus BM, Waller BF, Jones M, Epstein SE, Roberts WC: The case for preoperative coronary angiography in patients with tetralogy of Fallot and other complex congenital heart disease. *Am Heart J* 1982;103:451–456
14. Richardson JV, Doty DB, Rossi NP, Ehrenhaft JL: The spectrum of anomalies of aortopulmonary septation. *J Thorax Cardiovasc Surg* 1979;1:21–27
15. Bogers AJJC, Gittenberg-de Groot AC, Poelman RE, Peault BM, Huysmans HA: Development of the origin of the coronary arteries, a matter of ingrowth outgrowth? *Anat Embryol* 1989;180(5):437–441
16. Bortolotti U, Casarotto D, Betti D, De Mozzi PL, Stritoni P, Cevese PG: Anomalous origin of the right coronary artery from the main pulmonary artery. *Eur J Cardiol* 1978;7(5–6):451–455
17. Screenivasan VV, Jacobstein MD: Origin of the right coronary artery from the pulmonary trunk. *Am J Cardiol* 1992;69(17):1513–1514
18. Wald S, Stonecipher K, Baldwin BJ, Nutter DO: Anomalous origin of the right coronary artery from the pulmonary artery. *Am J Cardiol* 1971;27:677–681
19. Worsham C, Sanders SP, Burger BM: Origin of the right coronary artery from pulmonary trunk: Diagnosis by 2-D echocardiography. *Am J Cardiol* 1985;55:232–233
20. Eugster GS, Olivia PB: Anomalous origin of the right coronary artery from the pulmonary artery. *Chest* 1973;63,2:294–296
21. Lerberg DB, Ogden JA, Zuberbuhler JR, Bahson HT: Anomalous origin of the right coronary artery from the pulmonary artery. *Ann Thorac Surg* 1979;27:87–94
22. Huang TY, Hsueh Y, Tsung SH: Endocardial fibroelastosis and myocardial calcification secondary to anomalous right coronary artery arising from the pulmonary trunk. *Hum Pathol* 1985;16:959–960
23. Tingelstad JB, Lower RR, Eldredge WJ: Anomalous origin of the right coronary artery from the main pulmonary artery. *Am J Cardiol* 1972;30:670–673