The Turkish Journal of Pediatrics 1997; 39: 137-141

NON – INVASIVE DIAGNOSIS AND MANAGEMENT OF CORONARY ARTERIOVENOUS FISTULA*

A Case Report

Adnan Akçoral MD[**], *Vedide Tavlı MD*[***], *Meral Kozan MD*[***]
Oktay Ergene MD[****], *Talat Tavlı MD*[*****], *Nurettin Ünal MD*[***]

SUMMARY: Akçoral A, Tavlı V, Kozan M, Ergene O, Tavlı T, Ünal N. (Departments of Pediatric Cardiology and Cardiology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey). Noninvasive diagnosis and management of coronary arteriovenous fistula. A case report. Turk J Pediatr 1997; 39: 137-141.

Coronary arteriovenous fistulas are rare anomalies resulting in abnormal communication between the coronary artery and any chamber of the heart. An asymptomatic patient was referred for evaluation of her murmur. Two-dimensional and color Doppler echocardiographic evaluation revealed an enlarged left main coronary artery. A retrograde, eccentric small jet was found within the right ventricular outflow tract at the pulmonary artery valvular level allowing us to detect the entrance site of the fistula. The diagnosis was confirmed by cardiac catheterization and angiocardiography. Although our case was asymptomatic, the decision to perform cardiac surgery was made because of the aneurysmatic appearance of the left coronary artery. In our opinion, visualization of coronary arteries by two-dimensional echocardiography, together with additional information obtained from the Doppler examination, provides an excellent technique for the noninvasive diagnosis of coronary artery fistula. *Key words: coronary arteriovenous fistula, echocardiography.*

Coronary artery anomalies may be classified into three major groups, including anomalies with a shunt, anomalies without a shunt and congenital aneurysms and ectasia. Coronary artery fistulas are abnormal coronary artery communications that may involve any chamber and either or all coronary arteries[1]. In large series, the incidence has been reported to be less than 0.2 percent[2]. More than 90 percent of the communications are to the right side of the heart. Occasionally, both coronary arteries may participate in the fistula. Large shunts are particularly prevalent when the fistula terminates in the atrial chamber[3].

Case Report

An eight-year-old, asymptomatic girl was referred to our outpatient clinic for a murmur. Her growth parameters were at the 50th percentile. Her blood pressure and heart rate were normal, but a continuous murmur was heard at the 3rd left

* From the Departments of Pediatric Cardiology and Cardiology, Dokuz Eylül University Faculty of Medicine, İzmir.

** Professor of Pediatrics, and Pediatric Cardiologist, Dokuz Eylül University Faculty of Medicine.

*** Fellow in Pediatric Cardiology, Dokuz Eylül University Faculty of Medicine.

**** Associate Professor of Cardiology, Dokuz Eylül University Faculty of Medicine.

***** Assistant Professor of Cardiology, Dokuz Eylül University Faculty of Medicine.

intercostal space. Her EKG and telecardiography were normal. Segmental echocardiographic examination was obtained with Acuson-128 and 5 and 7 MHz transducers. The coronary artery anatomy was examined mainly in the parasternal short-axis view with the use of high-frequency transducers. In the parasternal short-axis plane, the aortic valve was seen to be flanked by the tricuspid valve on the left, and by the right ventricular outflow tract and pulmonary artery on the right (Fig. 1). Two-dimensional and Doppler echocardiographic evaluation revealed an enlarged left main coronary artery and a small color jet streaming retrograde mainly in diastole at the pravalvular level within the right ventricular outflow tract. The latter finding was considered as the site of the fistulous connection as seen in Figure 1. Right and left heart catheterization was performed to evaluate the magnitude of shunting and to demonstrate the coronary arteries selectively. Oxygen saturation did not show a significant step-up in the right ventricular outflow tract or the main pulmonary artery. Left- and right-sided pressures were normal. Cineangiographic films were obtained using Toshiba DF-P50 at 50 frames/sec with the DVM mode at 256 x 516 pixel density. Aortic root and selective left and right coronary artery injections showed a tortuous, ectasic main left coronary artery fistulating into the right ventricular outflow tract at the paravalvular level opacifying the main pulmonary artery (Fig. 2). The right coronary artery was normal.

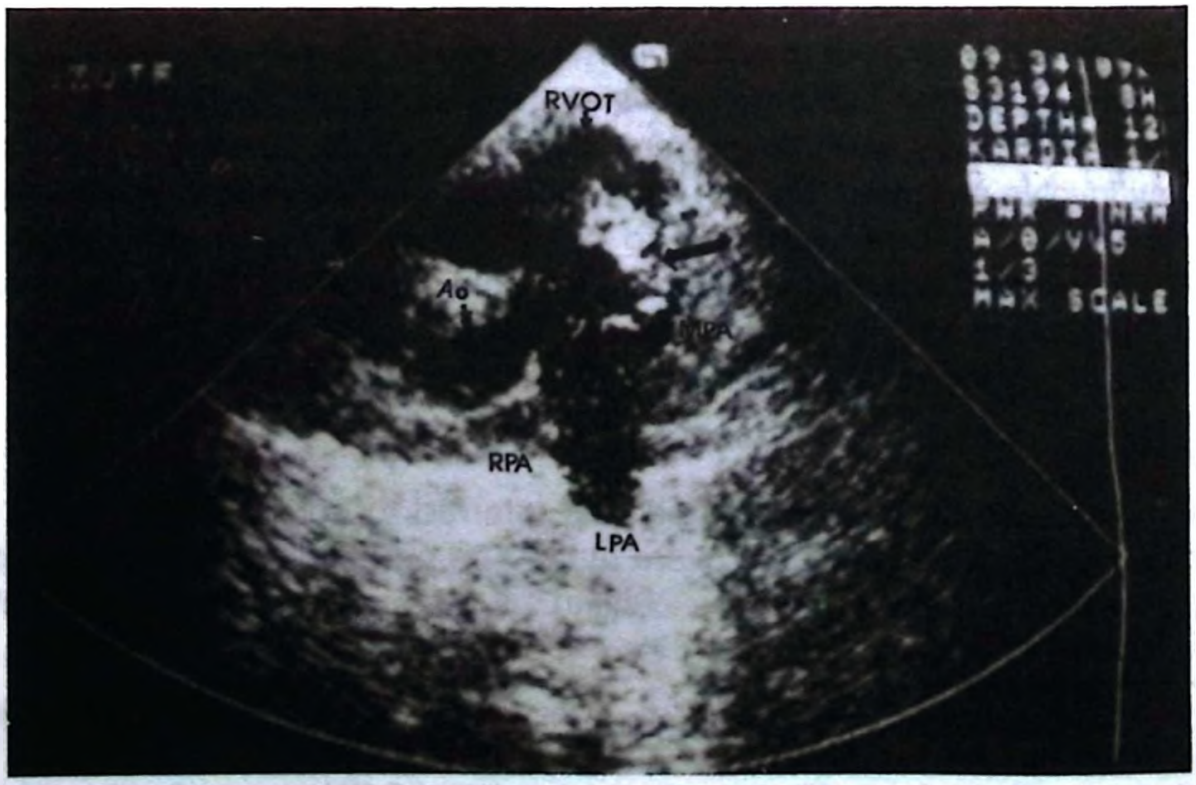

Fig. 1: Doppler color flow imaging of the left coronary artery fistulating into the right ventricular outflow tract in the parasternal short-axis view. The large arrow indicates the site of entrance of the fistula, producing a yellow aliasing jet area. Ao: aorta, MPA: main pulmonary artery. LPA: left pulmonary artery, RPA: right pulmonary artery, RVOT: right ventricular outflow tract.

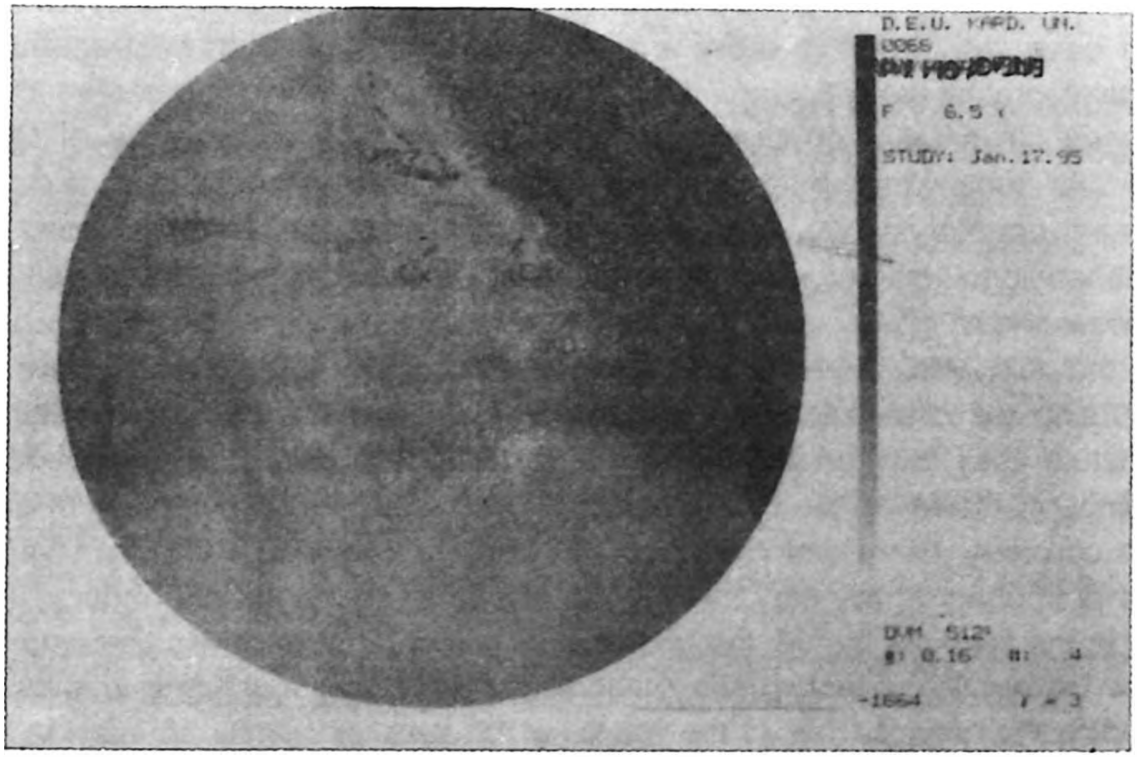

Fig. 2: Selective left coronary artery injection in the 45-degree, right anterior oblique projection shows an ectasic left main coronary artery giving rise to a thin segment which opacifies the pulmonary artery. The large arrow indicates the entrance site of the fistula into the right ventricular outflow tract at the pravalvular level. The fistulous jet is outlined with black dots. Cx: circumflex branch of the left coronary artery (LCA), LAD: left anterior descending branch of the LCA, MPA: main pulmonary artery. The large arrow indicates the entrance site.

Discussion

Coronary artery fistulas are rare anomalies. The incidence in our department was 1/4000 between July 1991 and January 1995. The incidence in large adult series has been reported at 0.2 percent[2].

The differential diagnosis in an acyanotic patient with a continuous murmur includes patent ductus arteriosus (PDA), aorto-pulmonary window, ventricular septal defect with aortic insufficiency, arterio-venous malformation of the chest wall or lung, and coronary artery fistula. The location of the maximum intensity of the continuous murmur gives a clue as to the appropriate clinical diagnosis[3]. In our case, the murmur was best heard at the left 3rd intercostal space, excluding the possibility of a PDA. Echocardiography and angiocardiography clearly differentiate such diagnoses.

The coronary artery anatomy may be especially important in certain forms of complex congenital heart disease, such as tetralogy of Fallot and transposition of the great arteries. Coronary artery anomalies have been reported to be detected by echocardiography[4,5]. Trans-esophageal echocardiography may be necessary

in older and overweight people in order to obtain efficient coronary artery imaging. In our case, the coronary artery anatomy was best deliniated by transthoracic echocardiography using 5 and 7 MHz transducers mainly in the parasternal short-axis view. In smaller children, the subcostal four-chamber view of the left ventricular outflow tract may be an adjunctive plane for visualization of portions of the left coronary artery. A coronary artery fistula should be suspected when two-dimensional imaging of the main coronary arteries in the parasternal short-axis view shows one coronary artery dilated while the other coronary artery is of normal size. Multiple two-dimensional echocardiographic planes were used in coursing the dilated left coronary artery in the parasternal short-axis plane. The fistula itself may be difficult to image despite a very carefully performed examination. However, the two-dimensional echocardiographic image of dilation of the coronary artery and detection of turbulent flow within the right ventricle or the pulmonary artery may identify the site of the fistulous connection. If flow through the fistula is heavy, the chamber or vessel that receives the fistula can be seen to be dilated. In our case, meticulous color Doppler imaging was essential for finding the entrance site of the fistula, which was very small as seen in Figure 1. The diagnosis was confirmed by angiocardiography, which showed prompt opacification of the right ventricular outflow tract during aortic root and selective coronary arteriography. The echocardiographer must remember that in order to position the Doppler beam in a parallel orientation to flow, an unusual position and angle may become necessary.

Coronary artery fistulas are generally reported as isolated lesions. Coronary artery-pulmonary artery communication may develop later in life in response to infection or profound cyanosis[6]. In the case presented, the left coronary artery was involved, which is relatively rare compared to right coronary artery fistulas[7]. Although the fistula in this case was thought to be congenital, the aneurysmatic appearance of the main coronary artery may suggest a previous Kawasaki disease episode that may have been misinterpreted as an allergic reaction, as learned from the patient's history.

Coronary arteriovenous fistulas have been reported to close spontaneously[8] Complications include infective endocarditis, myocardial ischemia, dysrhythmia and rupture[9-12]. Large fistulas may create hemodynamic overloading, resulting in heart failure. Aneurysmatic fistulas may gradually enlarge over time, compressing relatively softer cardiac structures or obstructing systemic or pulmonary venous return. Thrombosis and embolizations may occur. Myocardial ischemia may ensue due to the steal syndrome. Surgical treatment is recommended before any complications occur[13]. Clamping and ligating of the fistula is generally sufficient although, bypass operation may occasionally be necessary. Mortality is reported as zero to four percent[11, 13]. Microcoil embolization

and balloon occlusion of the fistula have recently been used successfully[14-16]. Our patient remains asymptomatic six months post-operatively.

In conclusion, although more than 50 percent of patients remain asymptomatic, coronary-arteriovenous fistulas require special attention in terms of possible complications if left untreated. Besides angiocardiography, which is the gold standard, echocardiography is considered to be a valuable diagnostic tool in the diagnosis of coronary artery anomalies in pediatric patients.

REFERENCES

1. Freedom RM, Culham JA, Moes CA. Angiocardiography of Congenital Heart Disease. Toronto: Collier MacMillan Inc; 1989: 405-427.
2. Yamanaka D, Habbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 1990; 21: 28-32.
3. Gillebert C, Van Hoof R, Van de Werf F, Piessens J, De Geest H. Coronary artery fistulas in an adult population. Eur Heart J 1986; 7: 437-443.
4. Sanders SP, Parness IA. Colan SD. Recognition of abnormal connections of coronary arteries with the use of Doppler color flow mapping. J Am Coll Cardiol 1989; 13: 922-926.
5. Douglas PS, Fiolkoski J, Berko B, Reichek N. Echocardiographic visualization of coronary artery anatomy in the adult. J Am Coll Cardiol 1988; 11: 565-571.
6. Haroituvan LM, Neill CA, Dorot JP. Pulmonary pseudofibrosis in cyanotic heart disease. A clinical syndrome mimicking tuberculosis in a patient with extreme pulmonary stenosis. Chest 1972; 62: 587-589.
7. Friedman WF. Congenital Heart disease in Infancy and Childhood. In: Braunwald E. (ed). Heart Disease (4th ed) Vol: 2. Philadelphia: W.B. Saunders; 1992: 917-918.
8. Hackett D, Hallidie-Smith KA. Spontaneous closure of coronary artery fistula. Br Heart J 1984; 52: 477-479.
9. Wilde P, Watt I. Congenital coronary artery fistulae: six new cases with a collective review. Clin Radiol 1980; 31: 301-311.
10. St. John Sutton MG, Miller GA, Kerr IH, Traill TA. Coronary artery steal via large coronary artery to bronchial artery anastomosis successfully treated by operation. Br Heart J 1980; 44: 460-463.
11. McNamara JJ, Gross RE. Congenital coronary artery fistula. Surgery 1969; 65: 59-62.
12. Haberman JH, Howard JL, Johnson ES, Rupture of coronary sinus with hemopericardium. Circulation 1963; 28: 1143-1146.
13. Meier B. Coronary occlusion after failed closure of coronaropulmonary fistula with detachable balloon. Cathet Cardiovasc Diagn 1989; 18: 237-239.
14. Nguyen K, Myler RK, Hieshima G, Ashraf M, Stertzer SH. Treatment of coronary artery stenosis and coronary arteriovenous fistula by interventional cardiology techniques. Cathet Cardiovasc Diagn 1989; 18: 240-243.
15. Strunk BL, Hieshima GB, Shafton EP. Treatment of congenital coronary arteriovenous malformations with micro-particle embolization. Cathet Cardiovasc Diagn 1991; 22: 133-136.
16. Reidy JF, Sowton E, Ross DN. Transcatheter occlusion of coronary to bronchial anastomosis by detachable balloon combined with coronary angioplasty in the same procedure. Br Heart J 1983; 49: 284-287.