The Turkish Journal of Pediatrics 1997; 39: 27-33

THE ANGIOCARDIOGRAPHIC ANALYSIS OF 73 PATIENTS WITH DOUBLE-OUTLET RIGHT VENTRICLE*

Ergün Çil MD[**], *Şencan Özme MD*[***], *Muhsin Saraçlar MD*[***]
Arman Bilgiç MD[***], *Süheyla Özkutlu MD*[***], *Sema Özer MD*[****]
Alpay Çeliker MD[****]

SUMMARY: Çil E, Özme Ş, Saraçlar M, Bilgiç A, Özkutlu S, Özer S, Çeliker A. (Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). The angiocardiographic analysis of 73 patients with double-outlet right ventricle. Turk J Pediatr 1997; 39: 27-33.

The purpose of this study was to determine the cardiac anatomy of patients with double-outlet right ventricle by angiocardiography. A total of 73 patients between the ages of one day and 11 years were examined. The aorta was on the right side of the pulmonary artery in 23 cases (32%), right anterior in 20 (27%) and right posterior in 17 cases (23%). Pulmonary stenosis was found in 53 patients (73%) and subaortic stenosis in six cases. Ventricular septal defect (VSD) was subaortic in 39 cases (52%), remote type in 17 (23%), doubly committed in 10 (13%), and subpulmonic in 9 (12%). Double VSD was noted in two patients. Pulmonary hypertension was more frequent in subpulmonic ventricular septal defect (78%). The most common associated anomalies were atrial septal defect (34%), anomalous coronary arteries (12%) and endocardial cushion defect (10%). Aortic root angiography was not satisfactory in half of the cases with coronary arterial anomaly. In conclusion, double-outlet right ventricle is a complex anomaly, all of whose cardiac features can be successfully demonstrated in detail by echocardiography and angiocardiography. However, in order to determine the anatomy of the coronary arteries, selective coronary angiography may be necessary in some patients. *Key words: double-outlet right ventricle, echocardiography, angiocardiography, cardiac catheterization, coronary artery.*

Double-outlet right ventricle (DORV) is an uncommon, congenital, cardiac anomaly in which the aorta and pulmonary artery reside predominantly above the morphologic right ventricle[1,2]. The natural history and clinical spectrum vary depending on the location of the ventricular septal defect (VSD), the presence or absence of pulmonary stenosis (PS), and other associated cardiac anomalies[3-7]. The feasibility of anatomic repair of DORV malformation has been reported with low mortality and morbidity rates[3,8,9]. To provide optimal surgical results, one must know the anatomy of the heart, great vessels and coronary arteries. The purpose of this study was to determine the entire cardiac anatomy by angiocardiography in 73 patients with the diagnosis of DORV.

* From the Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara.
** Assistant Professor of Pediatrics and Pediatric Cardiologist, Hacettepe University Faculty of Medicine.
*** Professor of Pediatrics and Pediatric Cardiologist, Hacettepe University Faculty of Medicine.
**** Associate Professor of Pediatrics and Pediatric Cardiologist, Hacettepe University Faculty of Medicine.

Material and Methods

The clinical records of the 234 patients who were diagnosed with DORV between January 1980 and September 1993 at Hacettepe Children's Hospital were reviewed. Of these patients, 73 cases with the following criteria were included the study: 1. only the patients whose clinical follow-up records were sufficient, 2. patients who had a satisfactory echocardiographic record, and 3. cases whose cineangiography was sufficient to depict all the cardiac anatomy.

For the purposes of this study, a patient was considered to have DORV when echocardiography, angiocardiography or direct inspection at operation demonstrated the origin of both great arteries to be the morphologic right ventricle, or the origin of one great artery and most of the other from the right ventricle with discontinuity of a great artery and mitral valves and with a bilateral subarterial conus[3, 10-12]. The diagnosis of DORV was made in all patients by both echocardiography and angiocardiography, and was confirmed intraoperatively in controversial cases. The categorization into morphologic subsets was made according to the commitment of the VSD to the great arteries as described by Lev et al.[2]. The relation of the great arteries was classified as in the study of Sridaromont et al.[1].

In all patients, the following parameters were evaluated: a) the position of the great arteries with respect to each other at the valvar level, b) the relationship of the ventricular septal defect and semilunar valves, c) the presence of subarterial stenosis, d) the anatomy of the coronary arteries, and e) the presence of associated defects.

Results

Seventy-three cases (45 boys and 28 girls) with the defined criteria were included the study. The ages of the patients at the time of diagnosis ranged from one day to 11 years (mean 2.8 ± 2.7 years, median 1.2 years). The mean age of the patients was 1.1 years at the time of palliative surgery and 4.5 years at corrective surgery.

The position of the semilunar valves with respect to each other is shown on Table I. The pulmonary outflow tract was devoid of obtruction in only 20 patients, while 53 (73%) had various forms of pulmonary stenosis. The pulmonary stenosis was valvar in eight patients, subvalvar (or infundibular) in 34 (Fig. 1) and both valvar and subvalvar in 11. The aortic outflow tract was obstructed in six patients. All of them had mild stenosis due to subvalvar hypertrophy. The location of the VSD was subaortic in 39 patients (52%), remote type in 17 {inlet in 11 (Fig. 2) and muscular in 6}, doubly committed in 10 and subpulmonic in nine cases. Two cases had double VSD, both perimembranous, subaortic and muscular.

Table I: Relation of the Aorta to the Pulmonary Artery

Relation	No. of Cases	%
Aorta on the right side	60	82
Side-by-side	23	32
Anterior (dextromalposition)	20	27
Posterior (normal relation)	17	23
Aorta on the left side	11	15
Anterior (levomalposition)	9	12
Side-by-side	2	3
Aorta in front of the pulmonary artery	2	3

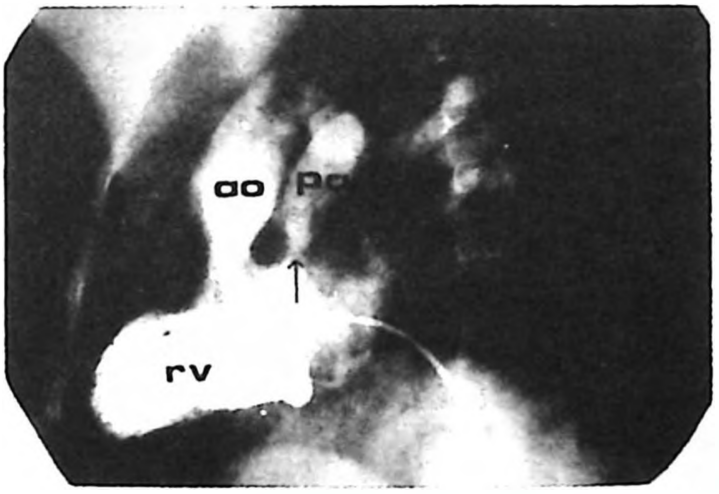

Fig. 1: Right ventriculagraphy (in lateral position) in a patient with double-outlet right ventricle. Aorta is anterior to the pulmonary artery and there is subvalvar pulmonary stenosis (arrow). ao: aorta, pa: pulmonary artery, rv: right ventricle.

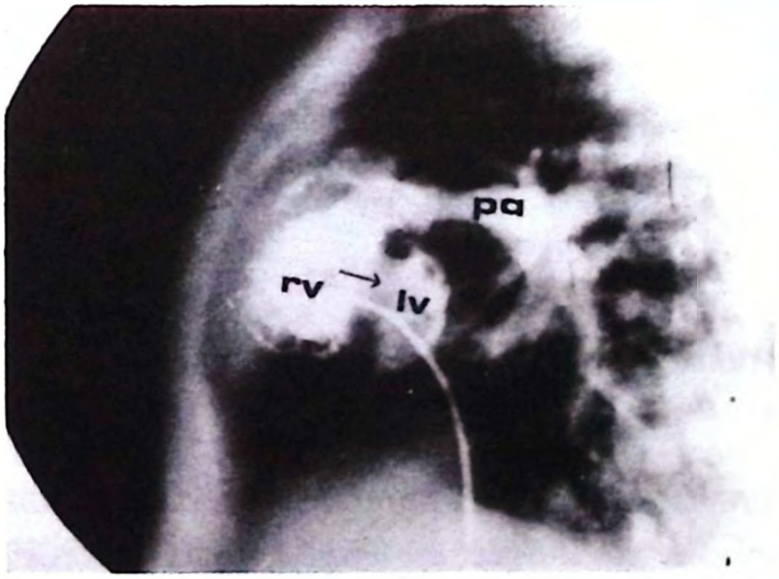

Fig. 2: The remote type (inlet muscular) of ventricular septal defect on right ventriculography (in lateral postion). lv: left ventricle, pa: pulmonary artery, rv: right ventricle, arrow: ventricular septal defect.

Pulmonary hypertension (PH) was present in 20 patients, and was severe in 14 cases. In the patients with PH, VSD was subpulmonic in seven cases, subaortic in five, remote type in four and doubly committed in four. The occurrence rate of pH according to the types of VSD was 78 percent in subpulmonic, 40 percent in doubly committed, 18 percent in remote type and 14 percent in subaortic (Table II).

Table II: The Rate of Occurrence of Pulmonary Hypertension by Type of VSD

VSD Type	PH/Patients	Rate (%)
Subpulmonic	7/9	78
Doubly committed	4/10	40
Remote type	4/17	18
Subaortic	5/39	13
Total	20/75*	27

* Double VSD was present in two patients. PH: pulmonary hypertension. VSD: ventricular septal defect.

Associated anomalies: All of the patients had at least one VSD. Atrial septal defect was the second-most common associated anomaly after VSD, while 18 patients had an ostium secundum defect. In addition, four paients had common atrium and three had ostium primum defect as part of the endocardial cushion defect. In six patients, a common atrioventricular valve was present. Patent ductus arteriosus was also present in seven cases. The position of the ventricles was abnormal in five patients: in four of them, the ventricles were in the side-by-side position (Fig. 3), and in one, they were superior-and-inferior. Anomalies of the coronary arteries were found in nine patients (12%). In five of them, the left anterior descending artery was originated from the right coronary artery. A single origin of the coronary arterial system (left) was present in two cases, with the right coronary artery coursed immediately beneath the pulmonary annulus; in two cases with levomalposition of the aorta, the right coronary artery coursed inferior to the pulmonary annulus.

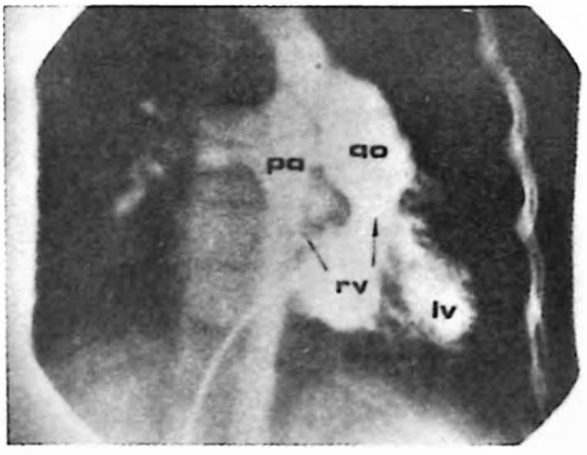

Fig. 3: Subaortic ventricular septal defect and bilateral subarterial conus (arrows) in a case with double-outlet right ventricle. note the side-by-side position of the ventricles. (Right ventriculography was performed in antero-posterior position).
ao: aorta, lv: left ventricle, pa: pulmonary artery, rv: right ventricle.

Discussion

Double-outlet right ventricle is a rare, congenital, cardiac anomaly that includes a diverse group of malformations sharing the common feature that both great arteries arise primarily from the right ventricle. The overall outlook for patients with DORV has improved greatly since the improvement of corrective procedures. However, surgical repair of DORV is complicated by the great variability in intracardiac and extracardiac morphology evidenced by these hearts. Complete correction of DORV mainly depends on the complexity of the cardiac anatomy. It is essential to obtain full detail of intracardiac anatomy and extracardiac anomalies before corrective surgery. Most of the previous series of cases with DORV have been reported by cardiac pathologists or cardiac surgeons, with pathologists tending to see patients with associated severe abnormalities, and surgeons tending to see patients free of such lesions[13]. Therefore, we analyzed the angiocardiographic data on all patients with DORV with or without surgical treatment.

In the present study, the median age of the patients (1.2 years) was greater than that in similar studies, since the patients from underdeveloped, rural regions of this country are generally older. In all cases, both great arteries were observed to originate predominantly from the morphologic right ventricle. In 82 percent of patients, the aorta was to the right of the pulmonary artery. The aorta and pulmonary artery were located side-by-side in 23 patients (32%); the side-by-side relation of the great arteries was also most common in previous studies[1, 5, 6, 11]. Dextromalposition (aorta on the right and in front of the pulmonary artery) was the second-most frequent type. The aortic levoposition was relatively uncommon, seen only in 15 percent of patients. The relation of the great arteries in the present study was nearly the same as in previous studies[1, 2, 5, 11]. However the location of the aorta in front of the pulmonary artery or to the left of the pulmonary artery (reverse side-by-side) was extremely uncommon in the medical literature[1, 6, 10, 11]. In the present study, the aorta was in front of the pulmonary artery in two patients and was on the left of the pulmonary artery (reverse side-by-side) in two patients.

The position of the great arteries must be defined clearly before corrective surgery because reconstruction of the pulmonary outflow tract is complicated when the pulmonary artery is behind the aorta. The location of the VSD is also important in patients with DORV. Remote-type (noncommitted) VSD and multiple VSDs in particular must be determined before surgery. Recognition of these types of defects is extremely important for the performance of a safe and complete repair. In the present study, noncommitted VSD was more common (23%) than in most of the previous studies. Judson et al.[11] reported three patients with remote-type VSD out of 62 patients (5%). The rate of remote-type VSD was 19 percent in the study by Macartney et al.[5], 17 percent in that of Hagler et al.[10], 10 percent in that of Sondheimer et al.[13] and 20 percent in that of Kirklin et al.[3].

Pulmonary hypertension is an anticipated outcome in patients with DORV without PS. Sondheimer et al.[13] reported that PS was present in 39 perecent of their patients with DORV. In the study of Judson et al.[11], PS was present in 74 percent of cases. The occurrence rate of PS was 61 percent in the study by Luber et al.[4]. In the present study PS was diagnosed in 73 percent of patients, and the remaining patients had PH of various severities. When the patients with PH were classified according to the site of VSD, it was observed that the majority of the VSDs were the subpulmonic type. The incidene of PH in the subpulmonic type of VSD was 78 percent; however, this rate was 40 percent in doubly committed VSD, and 13 percent in subaortic VSD. Therefore, early surgical treatment is compulsory in the subpulmonic type of VSD where the development of PH was extremely frequent.

The coronary arterial anatomy was drastically important in patients with DORV for both the arterial switch operation and pulmonary outflow reconstruction. There have been few studies examining the coronary anatomy in patients with DORV. Judson et al.[10] and Stellin et al.[14] reported high prevalences of anomalous coronary artery (28% and 22%, respectively); however, Luber et al.[4] reported only one patient (2%) with a coronary arterial anomaly out of a total of 57 cases. In the study presented, anomalous anatomy of the coronary arteries was determined in 12 percent of the patients. In all of these patients PS was accompanied by DORV. In spite of cardiac catheterization and angiography being performed in these patients, four of the coronary arterial anomalies were not diagnosed pre-operatively. Retrospectively it was perceived that aortic root angiography could not demonstrate the full coronary arterial anatomy in some patients because of the relatively inadequate amount of contrast material in the enormously enlarged aortic root. Therefore selective coronary angiography may be necessary in some patients.

In conclusion, double-outlet right ventricle is a complex anomaly in which all the cardiac features could be successfully demonstrated in detail by echocardiography and angiocardiography. However, to determine the anatomy of coronary arteries correctly, selective coronary angiography may be required in some patients.

REFERENCES

1. Sridaromont S, Ritter DG, Feldt RH, Davis GD, Edwards JE. Double-outlet right ventricle. Anatomic and angiocardiographic correlations. Mayo Clin Proc 1978; 53: 555-577.
2. Lev M, Bharat S, Meng CCL, Liberthson RR, Paul MH, Idriss F. A concept of double-outlet right ventricle. J Thorac Cardiovasc Surg 1972; 64: 271-281.
3. Kirklin JW, Pacifico AD, Blackstone EH, Kirklin JK, Bargeron LMJr. Current risks and protocols for operations for double-outlet right ventricle. Derivation from an 18 year experience. J Thorac Cardiovasc Surg 1986; 92: 913-930.
4. Luber JM, Castaneda AR, Lang P, Norwood WI. Repair of double-outlet right ventricle: early and late results. Circulation 1983; 68 (3 pt 2): II 144-147.
5. Macartney FJ, Rigby ML, Anderson RH, Stark J, Silverman NH. Double outlet right ventricle. Cross sectional echocardiographic findings, their anatomical explanation, and surgical relevance. Br Heart J 1984; 52: 164-177.

6. Piccoli G, Pacifico AD, Kirklin JW, Blackstone EH, Kirklin JK, Bargeron LMJr. Changing results and concepts in the surgical treatment of double-outlet right ventricle: analysis of 137 operations in 126 patients. Am J Cardiol 1983; 52: 549-554.
7. Serraf A, Lacour-Gayet F, Bruniaux J et al. Anatomic repair of Taussig-Bing hearts. Circulation 1991; 84 (5 Suppl): III 200-205.
8. Battistessa S, Soto B. Double outlet right ventricle with discordant atrioventricular connexion: an angiographic analysis of 19 cases. Int J Cardiol 1990; 27: 253-263.
9. Brawn WJ. Early results for anatomic correction of transposition of the great arteries and for double-outlet right ventricle with subpulmonary ventricular septal defect. J Thorac Cardiovasc Surg 1988; 95: 230-238.
10. Hagler DJ, Tajik AJ, Seward JB, Mair DD, Ritter DG. Double-outlet right ventricle: wide-angle two-dimensional echocardiographic observations. Circulation 1981; 63: 419-428.
11. Judson JP, Danielson GK, Puga FJ, Mair DD, McGoon DC. Double-outlet right ventricle. Surgical results. 1970-1980. J Thorac Cardiovasc Surg 1983; 85: 32-40.
12. Shen WK, Holmes DRJr, Porter CJ, McGoon DC, Ilstrup DM. Sudden death after repair of double-outlet right ventricle. Circulation 1990; 81: 128-136.
13. Sondheimer HM, Freedom RM, Olley PM. Double-outlet right ventricle: clinical spectrum and prognosis. Am J Cardiol 1977; 39: 709-714.
14. Stellin G, Ho SY, Anderson RH, Zuberbuhler JR, Siewers RD. The surgical anatomy of double-outlet right ventricle with concordant atrioventricular connection and noncommitted ventricular septal defect. J Thorac Cardiovasc Surg 1991; 102: 849-855.