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DOUBLE - OUTLET RIGHT VENTRICLE*

Morphological definition and surgical correction

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The rare entity called double-outlet right ventricle (DORV) in which both great vessels arise from the right ventricle has been recognized in one or other of its various forms since the last century[1-5]. During the past 30 years, the condition has been better understood and the detailed pathological findings have been accurately described[2,6,7]. Basically, the malformation is one in which both great vessels arise from the right ventricle and the only communication with the left ventricle is through a ventricular septal defect (VSD). This is true for the spectrum of malformations termed double-outlet right ventricle (DORV); correction of it by the tunnel-repair technique was first performed in 1956[6,8,9]. In this study we analyze our continuing surgical experience in this field and the surgical anatomy of patients with DORV.

Definition and Morphology: Double-outlet right ventricle (DORV) is one of the more complex congenital cardiac malformations in which there is an abnormal

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Key to Figures

Ao	: Aorta	CS	: Crista supraventricularis
PA	: Pulmonary artery	VSD	: Ventricular septal defect
PT	: Pulmonary trunk	DORV	: Double-outlet right ventricle
RA	: Right atrium	SVC	: Superior vena cava
RV	: Right ventricle	IVC	: Inferior vena cava
LA	: Left atrium		

ventriculo-arterial connection and both great arteries arise from the right ventricle. The only outlet from the left ventricle is through the ventricular septal defect (VSD)[9]. Several classifications for this entity have been offered[1,4,6,8,10-12], but we consider that a recent definition of DORV could be more suitable to surgeons and more meaningful in cases of surgical correction[6,8,10,13,14]. According to this definition, when 50% or more of each great artery (in relation to the ventricular septum) is arising from the right ventricle, the DORV is considered to be present, in situs solitus and atrioventricular concordance. Certain requirements, such as the presence of conal tissue between the semilunar and atrioventricular valves and above all, mitral aortic discontinuity, have been put forward as giving the precise anatomic definition of DORV. Recent morphological studies, however, have shown that mitral-aortic continuity is not always to be found[6,8,10,13,14]. Hearts with DORV (Figure 1) are classified into widely diverse groups in relation to

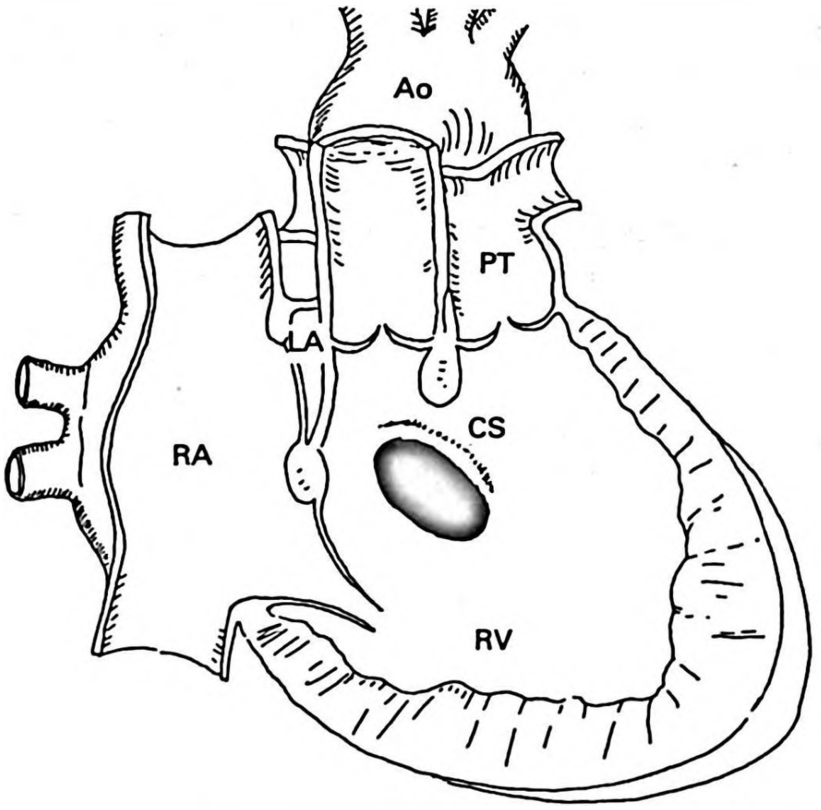

Figure 1

Showing the types of DORV according to the types and the position of VSD.

1 – a) DORV with aortic committed (subaortic) VSD.

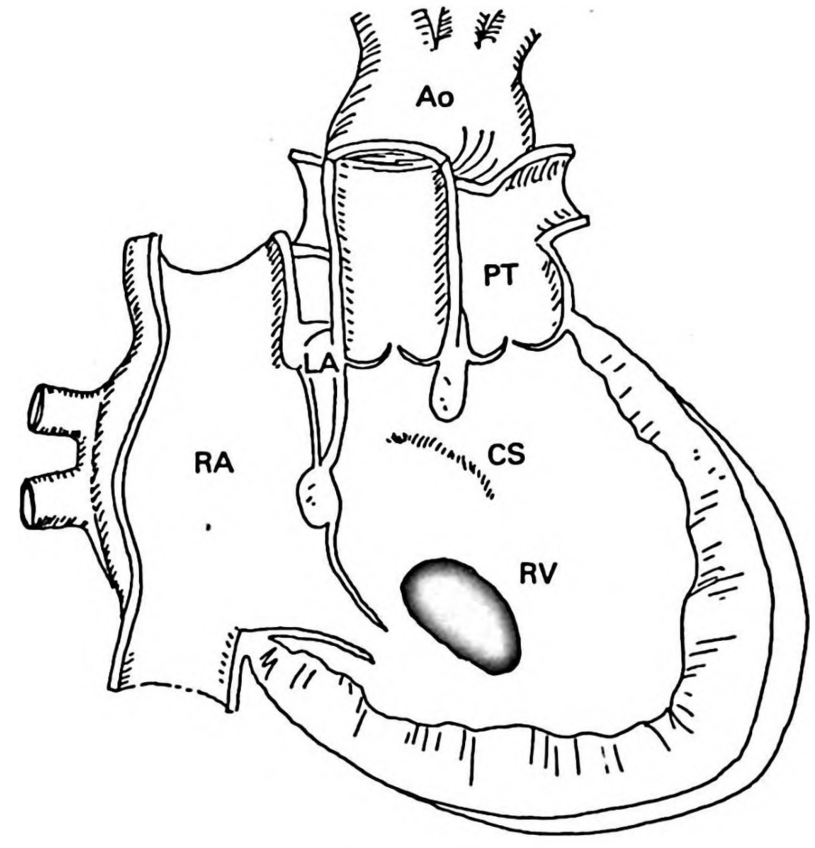

Figure 1 – b
DORV with non-committed VSD.

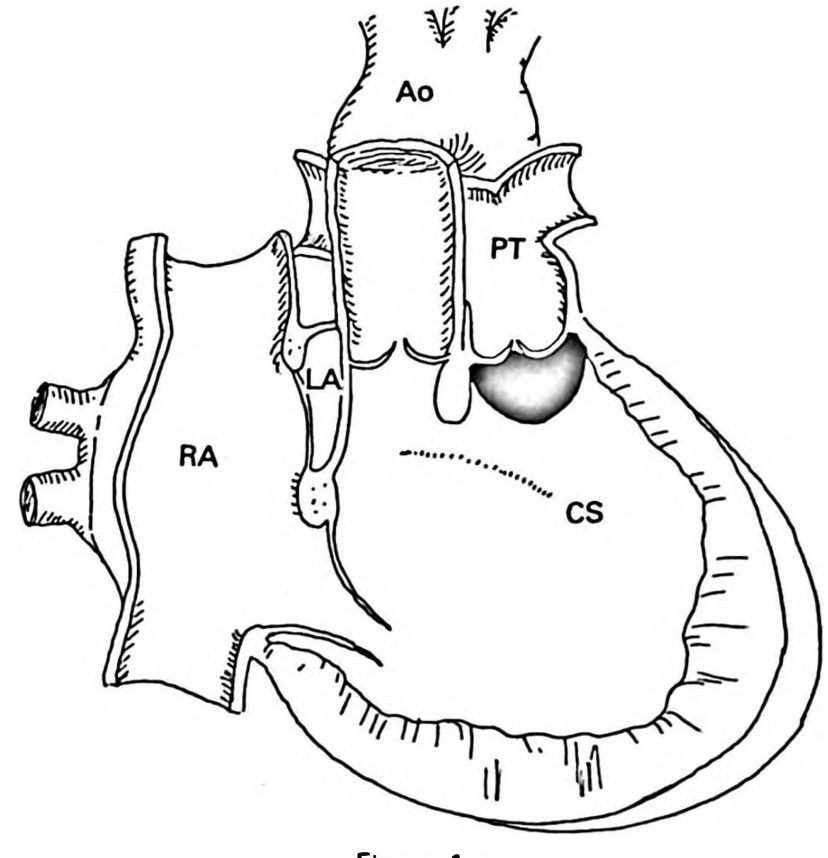

Figure 1 – c
DORV with subpulmonary VSD.

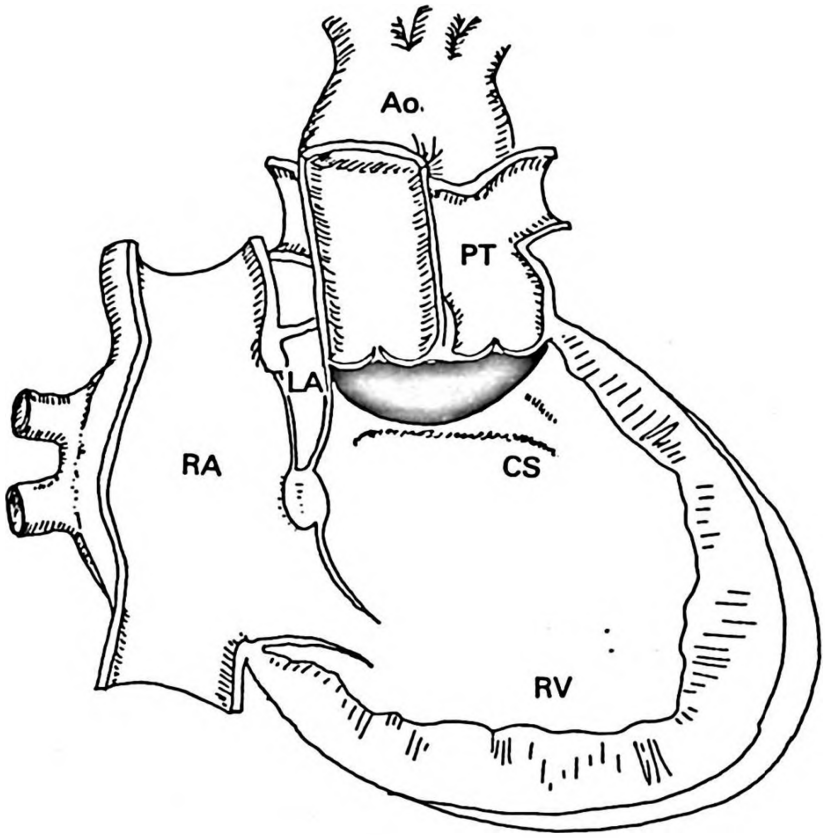

Figure 1 – d
DORV with doubly committed VSD.

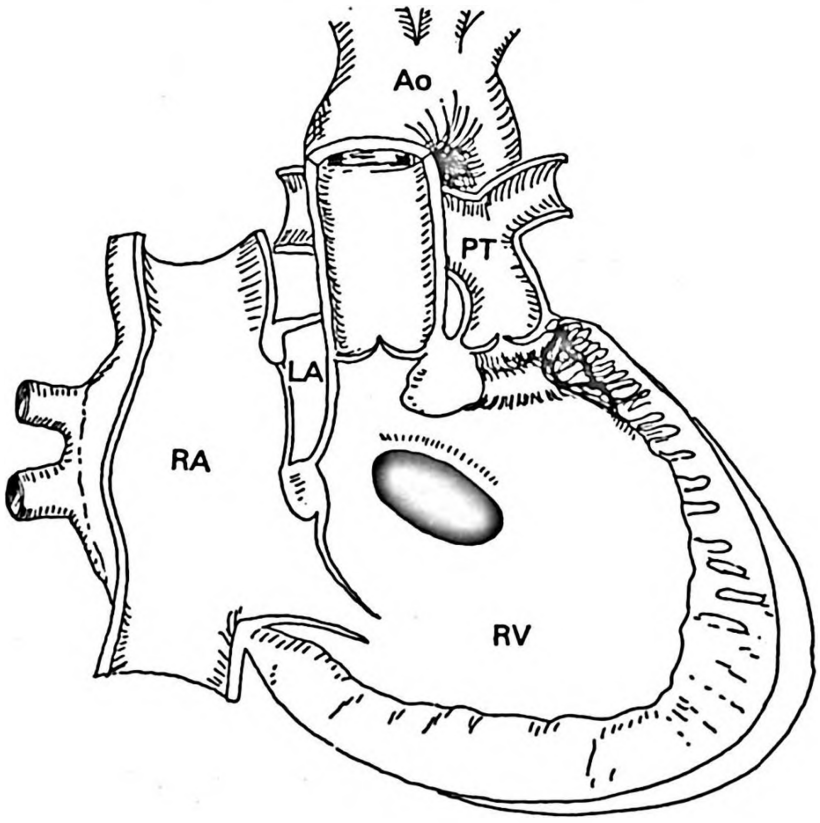

Figure 1 – e
DORV with aortic committed (subaortic) VSD and subaortic stenosis.

several associated factors[6,8,9,11,13]: 1. the types and positioning of the ventricular septal defects; 2. the position of the great arteries relative to themselves and to the VSD; 3. the presence or the absence of subpulmonary or subaortic stenosis; and 4. the presence of other significant anomalies. In the vast majority of cases the VSD lies between the two limbs of the trabeculoseptomarginalis and may be subaortic, subpulmonary or doubly committed[6,8,10,15,16]. Malinsertion and malposition of the infundibular septum determine the location of the VSD[4,8,17,18].

The VSD is considered subaortic (aortic committed) when it is immediately beneath the aortic valve or subaortic conus. Generally the VSD in this situation is perimembranous in location and up against the commissure between anterior and septal tricuspid leaflet. On occasion, such defects may be small and thereby restrict the outlet of the left ventricle.

The VSD is considered doubly committed when it is clearly immediately beneath both the aorta and the pulmonary artery. In such cases the aortic and pulmonary valves are almost contiguous with each other, and the VSD is very large. The VSD is considered to be noncommitted (or remote)[11] when it is adjacent neither to the aorta nor to the pulmonary artery. Such defects are in the trabecular or apical portion of the muscular septum, or beneath the septal leaflet of the tricuspid valve (so-called atrioventricular canal type of VSD)[6,8,9,12,13,17].

In a few patients with DORV the aorta is to the left and more or less side by side with the pulmonary artery (known as truncal inversion)[8].

Subpulmonary stenosis is created by infundibular obstruction. Valvular pulmonary stenosis is frequently combined with infundibular stenosis[9]. Subpulmonary stenosis, when present, may be of the tetralogy or nontetralogy type. The former is characterized by insertion of the infundibular septum anterosuperiorly to the superior limb of the trabeculoseptomarginalis, while the latter is not[16].

Subaortic stenosis may very occasionally be seen when the subaortic conus is present[8].

In the Taussig-Bing type of DORV[3,6], the aorta is usually anterior to the pulmonary artery, either directly so, or slightly to the right or to the left. The VSD is cephalad and anterior in the septum and related to the pulmonary artery (pulmonic committed VSD).

The diagnosis of DORV is appropriate whenever 50% or more of each of the great vessels is arising from the right ventricle[6,8,10,13,14]. Patients with tetralogy of Fallot-like malformations are considered to have DORV, if 80-90% or more of the aorta arises from the right ventricle. The two anomalies may occur independently or may coexist in the sarne heart. Cases of tetralogy in which only slightly more than 50% of the aorta arises from the right ventricle may be tetralogy with DORV. Those cases of DORV with tetralogy having subpulmonary stenosis in which

80-90% of the origin of the aorta is from the right ventricle, may be considered DORV with tetralogy[16].

Material and Methods

Between September, 1974 and December, 1983, 11 patients with concordant atrioventricular connection, and two ventricles, underwent complete intracardiac repair of DORV, and they form the basis for this study. Nine (81%) of these 11 patients had pulmonary stenosis. Ages ranged from four to eighteen years and the median age was 9.3 years. In five of our patients, echocardiographic examinations showed aortic-mitral discontinuity. Diagnostic and hemodynamic details are summarized in Table I.

TABLE I

Diagnostic Findings

Case no., Initials	Age in years	Morphologic description	Result
1 Ü.E.	4	DORV* with aortic committed VSD, without PS.	5-year F-U*, NYHA Class II
2 D.D.	9	DORV with aortic committed VSD, with moderate PS.	4-year F-U, NYHA Class I-II
3 S.U.	18	DORV with aortic committed VSD, with moderate PS.	Exitus in early postoperative period (A-V dissociation)
4 R.S.	18	DORV with restrictive, non-committed VSD, with pulmonary atresia.	Exitus in early postoperative period
5 M.U.	8	DORV with restrictive, aortic committed VSD, with moderate PS.	5-year F-U, NYHA Class II
6 A.I.	6	DORV with aortic committed VSD, with severe PS.	1-year F-U, NYHA Class I
7 M.D.	9	DORV with restrictive, non-committed VSD, with moderate PS.	Exitus in early postoperative period (Complete A-V Block)
8 F.A.	6	DORV with aortic committed VSD, without PS.	2-year F-U, NYHA Class I
9 Ö.A.	7	DORV with restrictive, aortic committed VSD with severe PS.	Exitus on the 28th day postoperatively
10 T.H.	9	DORV with aortic committed VSD, with moderate PS, with coronary artery anomaly	1-year F-U, NYHA Class I
11 M.Ö.	9	DORV with restrictive, aortic committed VSD, with mild PS.	3-month F-U, NYHA Class I

F-U : Follow-up.
DORV : Double-outlet right ventricle.
VSD : Ventricular septal defect.
PS : Pulmonary stenosis
NYHA : New York Heart Association.

Two patients (18%) with DORV and non-committed VSD had previously undergone Waterston-Cooley anastomosis. One of these had pulmonary atresia and a diminutive left ventricle.

The operations were carried out by means of median sternotomy, and a cardiopulmonary bypass was used. Until about 1978, the technique involved

systemic moderate hypothermia and anoxic cardiac arrest. From 1978 to 1983 this method was replaced by moderate whole body hypothermia, external cardiac cooling and cold K^+ cardioplegia for myocardial preservation.

Intraventricular repair of DORV was generally accomplished by the tunnel repair technique, a patch being placed so as to conduct left ventricular blood coming through the VSD under the patch and into the aorta[15] (Figure 2 a,b,c).

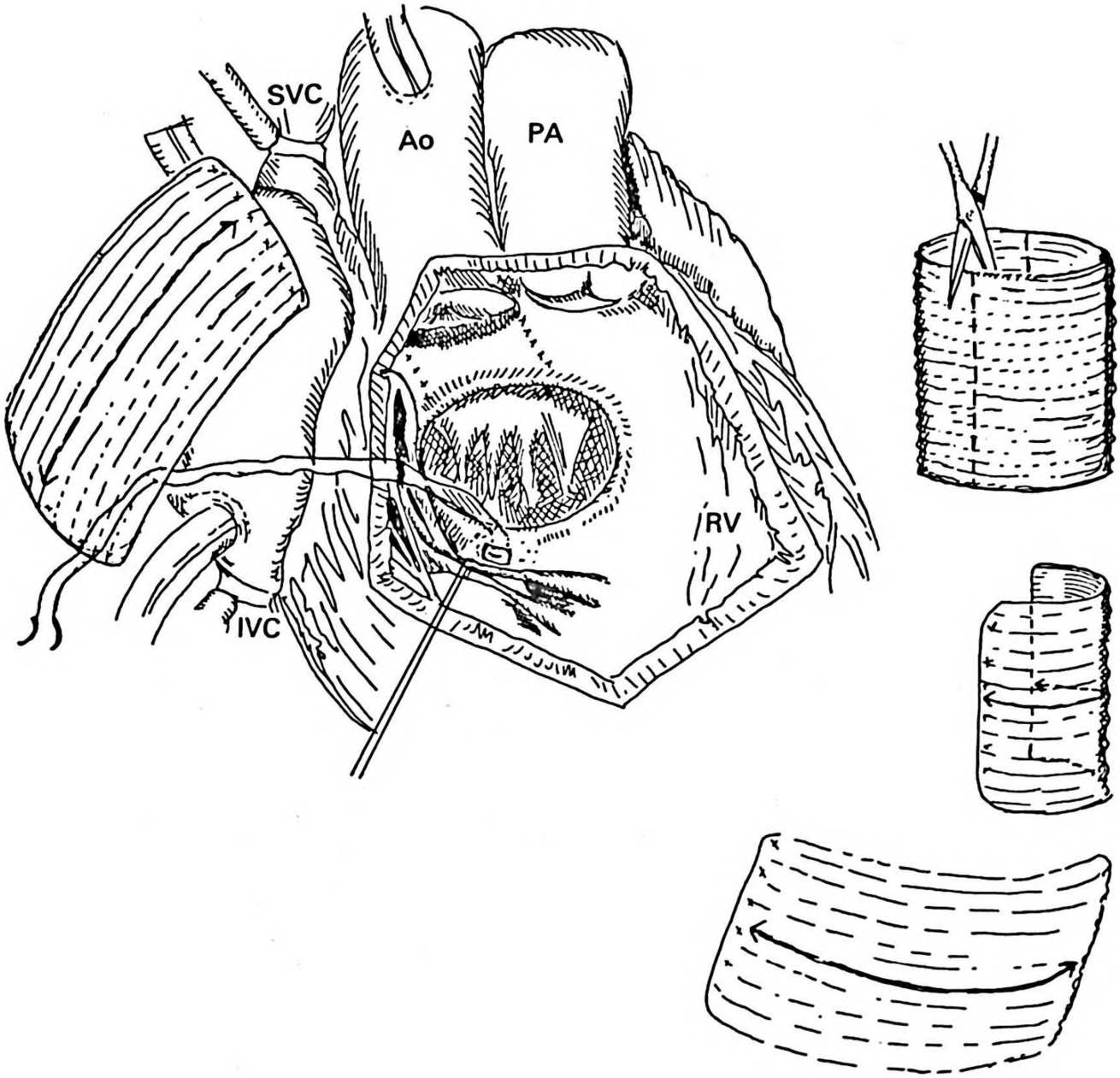

Figure 2

Showing repair of DORV with subaortic (aortic committed) VSD.

a) Showing the trimming and the insertion of the tubular dacron graft. The tubular dacron graft should be little larger than the ascending aorta. The length of the graft must be adjusted according to the distance between the VSD and the aortic annulus.

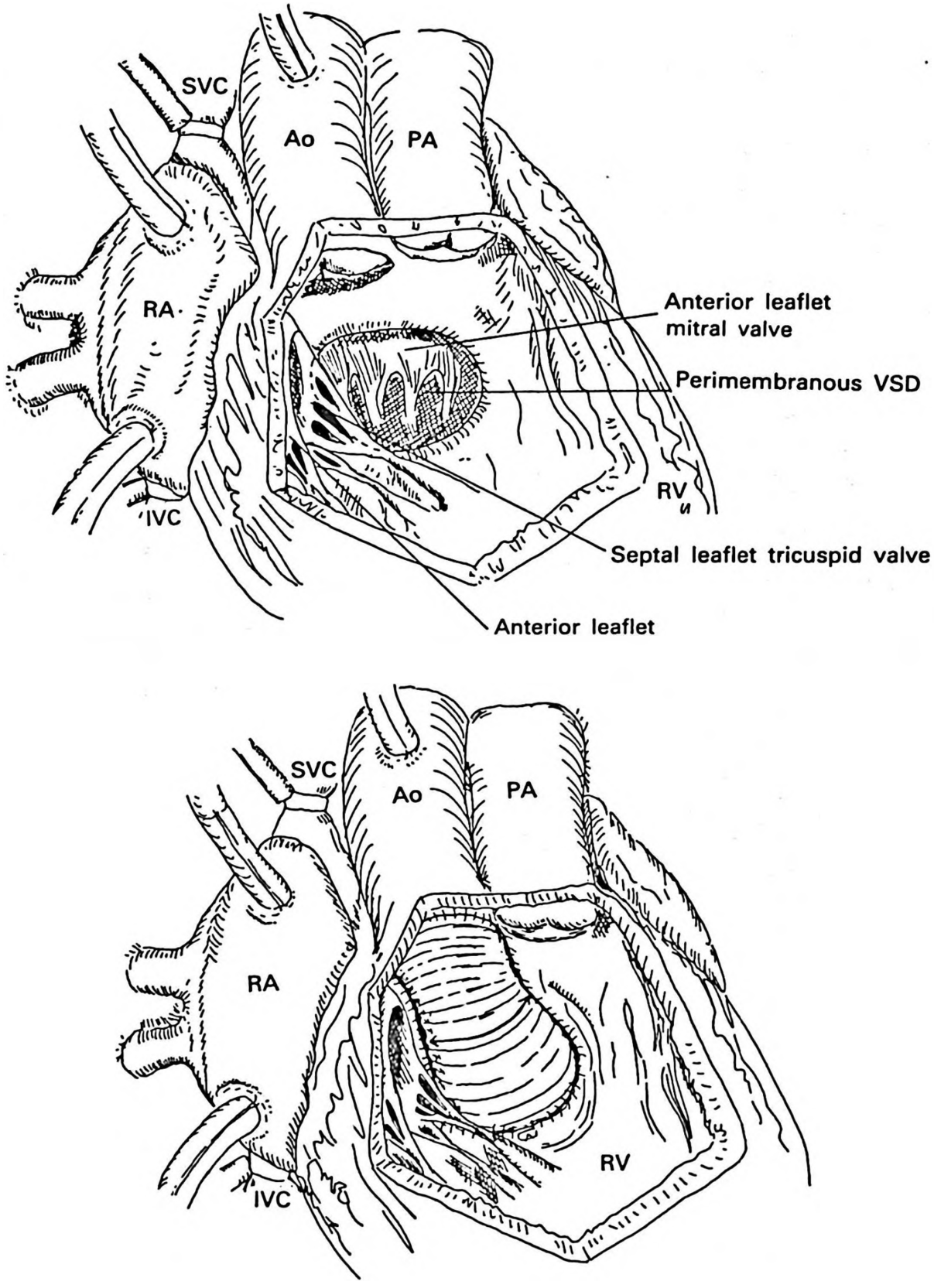

Figure 2

b and c) Showing the final situation of the tubular dacron graft with proper geometry. The final result is best visualized as a subaortic tunnel. The posterior third of this tunnel is formed by subaortic infundibulum, the other two-thirds by the dacron.

When the VSD was restrictive, it was surgically enlarged. In five (45%) of our cases, the VSD were restrictive, and these were surgically enlarged from the anterosuperior aspect (Figure 3). Two patients had non-committed and restrictive VSD. These were repaired but the patients subsequently died as a result of low cardiac output.

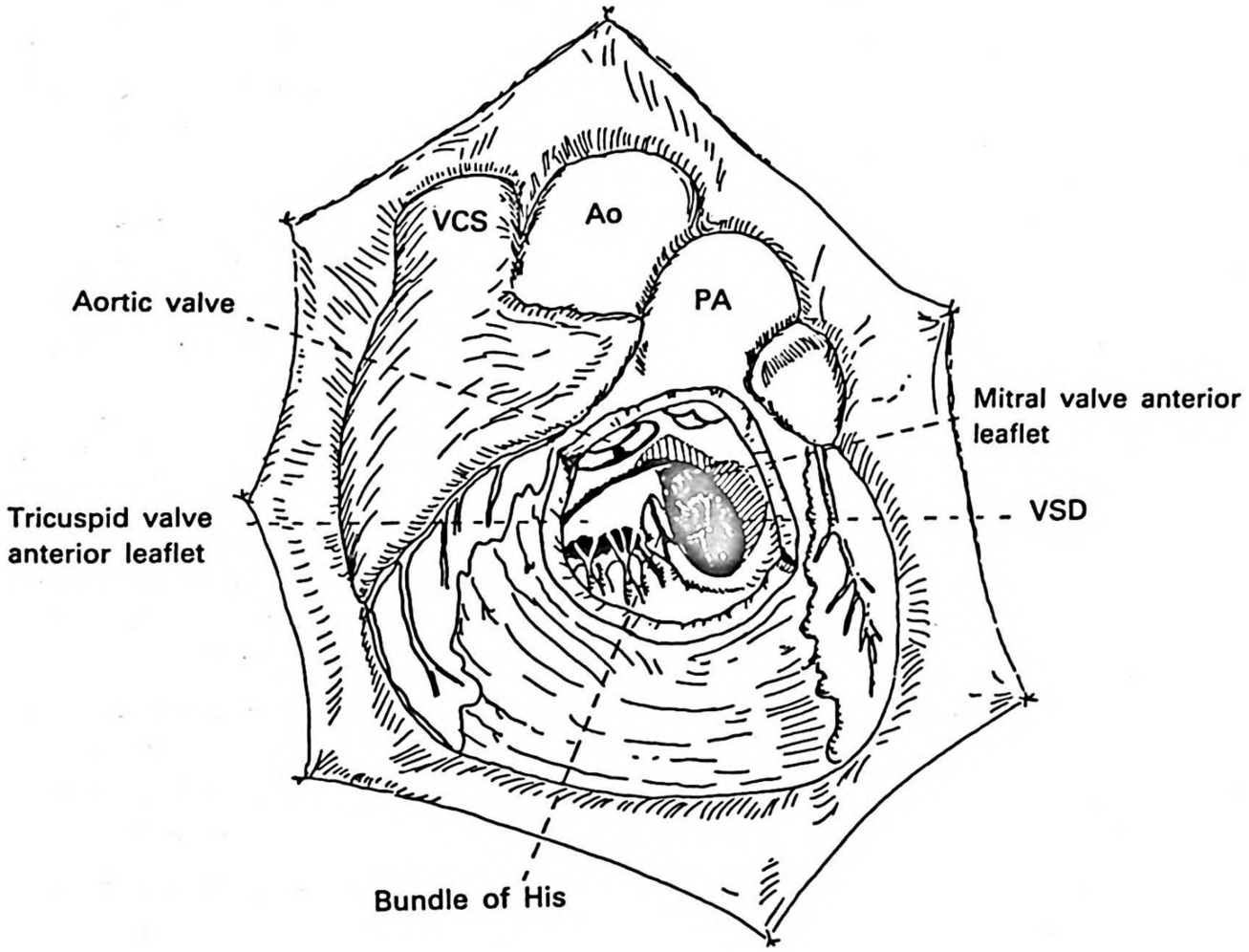

Figure 3

Showing the enlargement of restrictive VSD. The shadowed area shows the site of enlargement.

Pulmonary stenosis was corrected by infundibular resection and pulmonary valvotomy or by patch graft enlargement of the outflow tract or by a valved external conduit. We performed transannular patch enlargement in seven cases, infundibular resection and pulmonary valvotomy in three cases and a valved external conduit was used in one patient.

In seven of our cases, 100% of the aorta was arising from the right ventricle while in the other four patients more than 80% of the aorta was in the dextroposition.

Results

Three (27%) of the 11 patients operated on died from low cardiac output in the early postoperative period. Another died from mediastinitis and sepsis 28 days after the operation. In all of these patients there had been severe pulmonary stenosis. A valved external conduit had been used in one patient and three had undergone transannular patch graft enlargement. In two of the fatal cases there was non-committed VSD. This position of the VSD was an incremental risk factor. Complete heart block occurred in two (18%) of the patients. One of these had junctional rhythm after repair (operation in 1981) with a ventricular rate of 55 to 60 beats per minute, however, two years postoperatively, this patient still did not need a permanent pace-maker. Although we have inadequate data concerning the late postoperative course of those who survived, we do know that there have been no deaths in this group. Our overall mortality rate was 36%.

Discussion

By means of some recent morphological investigations, many important improvements in the scope of corrective surgery for cases of double-outlet right ventricle have been obtained. Today, when describing DORV,although there are still some conflicting opinions it is usual to emphasize that more than 50% of both of the great vessels are originating from the right ventricle[6,8,13,14]. This concept is comprehensive and useful from the point of view of surgical anatomy and with respect to the form of surgical procedure that will be chosen. The surgical correction is mainly concerned with the relations between the great vessels, the interventricular septum, and VSD localization. From a surgical point of view, the absence or the presence of a conal tissue between semilunar and atrioventricular valves (mitral-aortic discontinuity or continuity) is of no importance[6,13,14].

The main pointin the total correction of DORV is the creation of an intraventricular tunnel facilitating blood flow from the left ventricle to the aorta. While carrying out this procedure, the right ventricular outflow obstruction, whether present before or as a result of tunnel repair, should be corrected by means of infundibular resection, with replacement of an outflow patch, or an external conduit[12,13,18,19].

For intraventricular tunnel repair, the use of a tubular dacron graft or a dacron-teflon patch trimmed to the proper dimensions has been recommended[17]. In one of our patients, we used a tubular dacron graft, but we are of the opinion that a dacron-teflon patch trimmed to shape would have been sufficient[12] (Figure 2).

In the presence of subaortic or doubly committed VSD, intraventricular tunnel repair is relatively easy. The most important point in this procedure, is the reconstruction of the tunnel in such a way as not to cause left ventricular outflow obstruction[17,20].

If there is a restrictive VSD, it is enlarged from the anterosuperior edge. In these patients, the interventricular septum may be relatively thicker, therefore, to ensure sufficient enlargement, a relatively large muscle mass should be resected and the size of the VSD must at least equal the diameter of the aortic annulus[12,17]. We used this VSD enlargement technique on five of our patients and it brings no incremental risks (Figure 3).

In the patients with pulmonary stenosis, right ventricular outflow tract reconstruction is usually needed in addition to infundibular resection[12,13,17]. Replacement of a transannular patch may be sufficient in most of the cases[12]. Because of the difficulties involved, in placing the extracardiac conduits into the thoracic cavity and because of the degenerative and obstructive problems often encountered in long term follow-ups[21], we prefer to use the outflow patch system.

Generally in the Taussig-Bing group, one of these two treatment options must be selected: 1. Closure of the VSD so as to produce transposition of the great arteries and then the correction of it by using the Mustad or Senning technique. (If pulmonary stenosis is present, a valved external conduit from left ventricle to pulmonary artery can be used simultaneously), 2. Tunnel repair of the VSD so that the left ventricle ejects into the aorta and closure of the pulmonary artery with the placement of a valved external conduit between the right ventricle and the pulmonary arter. Otherwise, the Damus-Kaye-Stansel operation may be performed[3,6,11,17,19,20].

Because of the pulmonary vascular obstructive disease, patients without pulmonary stenosis must be corrected completely before one year of age[20,22-24]. Instances of severe pulmonary stenosis should be palliated by balloon septostomy (if the patient is under one month of age) and by systemic pulmonary shunt (for those in the first year of life). Definitive repair should be carried out at the age of 3 to 5 years[24-26].

According to recent studies, the overall mortality rate for cases with DORV has been ranging from 15% to 34%[12,13]. In our series, the overall mortality rate was 36%, or 27% if we exclude the case where death was the result of mediastinitis and sepsis.

The most important cause of mortality in the early postoperative period is the low cardiac output resulting from acute cardiac failure, especially in patients whose right ventricular outflow tracts were not effectively reconstructed. After an adequate right ventricular outflow tract reconstruction, the gradient between the pressures in the right ventricle and the pulmonary artery should be as low as possible[13,17].

We have few data about the long-term follow-up of our patients. But in the literature it is indicated that the arrhythmias could eventually lead to death.

Therefore patients who have received surgical treatment for DORV should be regarded as at risk for arrhythmia and the benefits to these patients of rapidly evolving modern techniques and regimens[27-30] should be kept in mind[13,27-30].

Summary

Eleven patients with double-outlet right ventricle (DORV) underwent total correction between September 1974 and December 1983. All of the patients had relatively uncomplicated DORV (sub-aortic or noncommitted VSD and with or without pulmonary stenosis). The intracardiac tunnel repair technique was performed in the majority of the patients and a valved external conduit was used in one. In this report, in addition to the presentation of our cases, morphological definition and surgical technique have been discussed in detail.

REFERENCES

1. Witham AC. Double outlet right ventricle. A partial transposition complex. *Am Heart J* 53:928, 1957.
2. Edwards WD, Wilcox WD, Danielson GK, Feldt RH. Postoperative false aneurysm of left ventricle and obstruction of left circumflex coronary artery complicating enlargement of restrictive ventricular septal defect in double-outlet right ventricle. *J Thorac Cardiovasc SURG* 80:141, 1980.
3. Lev M, Rimoldi HJ, Eckner FA, et al. The Taussig-Bing heart. Qualitative and quantitative anatomy. *Arch Pathol* 81:24, 1966.
4. Van Praagh R. What is the Taussig-Bing malformation? *Circulation* 38:445, 1968.
5. Anderson RH, Wilkinson JL. Arnold R, et al. Morphogenesis of bulboventricular malformations. II. Observations on malformed hearts. *Br Heart J* 36:948, 1974.
6. Lev M, Bharati S, Meng CC, et al. A concept of double-outlet right ventricle. *J Thorac Cardiovasc Surg* 64:271, 1972.
7. Bharati S, Kirklin JW, McAllister HA Jr, Lev M. The surgical anatomy of common atrioventricular orifice associated with tetralogy of Fallot, double-outlet right ventricle and complete regular transposition. *Circulation* 61:1142, 1980.
8. Wilcox BR, Ho SY, Macartney FJ, et al. Surgical anatomy of double-outlet right ventricle with situs solitus and atrioventricular concordance. *J Thorac Cardiovasc Surg* 82:405, 1981.
9. Moss AJ, Adams FH, Emmanouilides GJ (eds). *Heart Disease in Infants and Adolescents* (2nd ed). Baltimore: Williams & Wilkins, 1977, p 355.
10. Anderson RH, Shinebourne EA. *Paediatric Cardiology.* New York: Churchill Livingstone, 1978, p 36.
11. Zamora R, Moller JH, Edwards JE. Double-outlet right ventricle. Anatomic types and associated anomalies. *Chest* 68:672, 1976.
12. Judson JP, Danielson GK, Puga FJ, et al. Double-outlet right ventricle. Surgical results, 1970-1980. *J Thorac Cardiovasc Surg* 85:32, 1983.
13. Stewart S. Double-outlet right ventricle. A collective review with a surgical viewpoint. *J Thorac Cardiovasc Surg* 71:355, 1976.
14. Kirklin JW. *Advances in Cardiovascular Surgery.* New York: Grune and Stratton, 1973, p 57.
15. Kirklin JW, Harp RA, McGoon DC. Surgical treatment of origin of both vessels from right ventricle, including cases of pulmonary stenosis. *J Thorac Cardiovasc Surg* 48:1026, 1964.

16. Edwards WD. Double-outlet right ventricle and tetralogy of Fallot. Two distinct but not mutually exclusive entities. *J Thorac Cardiovasc Surg* 82:418, 1981.

17. Stewart RW, Kirklin JW, Pacifico AD, et al. Repair of double-outlet right ventricle. An analysis of 62 cases. *J Thorac Cardiovasc Surg* 78:502, 1972.

18. Sridaromont S, Ritter DG, Feldt RH, et al. Double-outlet right ventricle. Anatomic and angiocardiographic correlations. *Mayo Clin Proc* 53:555, 1978.

19. McGoon DC. Origin of both great vessels from the right ventricle. *Surg Clin N Amer* 41:1113, 1961.

20. Rocchini A, Rosenthal A, Castaneda AR, et al. Subaortic obstruction after the use of an intracardiac baffle to tunnel the left ventricle to the aorta. *Circulation* 54:957, 1976.

21. Ciaravella JM, McGoon DC, Danielson GK, et al. Experience with the extracardiac conduit *J Thorac Cardiovasc Surg* 78:920, 1979.

22. Engle MA, Steinberg I, Lukas DS, Goldberg HP. Acyanotic ventricular septal defect with both great vessels from the right ventricle. *Amer Heart J* 66:755, 1963.

23. Chaitman BR, Grondin CM, Théroux P, Bourassa MG. Late development of left ventricular outflow tract obstruction after repair of double-outlet right ventricle. *J Thorac Cardiovasc Surg* 72:265, 1976.

24. Harvey JC, Sondheimer HM, Williams WG, et al. Repair of double-outlet right ventricle. *J Thorac Cardiovasc Surg* 73:611, 1977.

25. Tandon R, Moller JH, Edwards JE. Communication of mitral valve with both ventricles associated with double outlet right ventricle. *Circulation* 48:904, 1973.

26. Kitamura N, Takao A, Ando M, et al. Taussig-Bing heart with mitral valve straddling: case reports and postmortem study. *Circulation* 49:761, 1974.

27. Deanfield JE, McKenna WJ, Hallidie-Smith KA. Detection of late arrhythmia and conduction disturbance after correction of tetralogy of Fallot. *Br Heart J* 44:248, 1980.

28. Steeg CN, Krongrad E, Davachi F, et al. Postoperative left anterior hemiblock and right bundle branch block following repair of tetralogy of Fallot. Clinical and etiologic considerations. *Circulation* 51:1026, 1975.

29. Wessel HU, Bastanier CK, Paul MH, et al. Prognostic significance of arrhythmia in tetralogy of Fallot after intracardiac repair. *Am J Cardiol* 46:843, 1980.

30. Garson A Jr, Gillette PC, Gutgesell HP, McNamara DG. Stress-induced ventricular arrhythmia after repair of tetralogy of Fallot. *Am J Cardiol* 46:1006, 1980.