Double Outlet Right Ventricle

A review of 16 cases with 10 necropsy specimens

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In so-called "double outlet right ventricle" both aorta and pulmonary trunk arise wholly from the right ventricle. This relatively uncommon abnormality is of interest anatomically and physiologically and has considerable surgical significance. The condition has been described by various authors including Witham (1957), Neufeld, DuShane, and Edwards (1961a), Neufeld *et al.* (1961b, 1962), and Morgan *et al.* (1962), with emphasis on the variations in patterns dependent on the presence or absence of significant pulmonary stenosis. Its embryology has been discussed by Grant (1962) and by Van Mierop and Wiglesworth (1963).

This paper describes a further 16 cases of double outlet right ventricle, 10 of which have come to necropsy. These cases illustrate the variations in anatomy that occur in this condition and the clinical syndromes associated with it.

SUBJECTS

Nine patients have been seen since 1960 in the Cardiac Investigatory Clinic of the Royal Children's Hospital, Melbourne, and at necropsy it has been shown that they have double outlet right ventricle. An additional specimen was obtained from the pathology files of the hospital. Double outlet right ventricle has been diagnosed by special investigation in 6 other surviving patients. Patients in whom the pulmonary trunk overrides a ventricular septal defect have been excluded.

The clinical features and other findings of the patients who died with double outlet right ventricle and who were subjected to necropsy will be described briefly. The material is arranged with regard to variations in anatomical relationship between the rings of the mitral, aortic, and pulmonary valves and the ventricular septal defect. Table I summarizes these findings. Details of patients in whom the diagnosis is based on investigatory

findings alone are given in Table II. The investigatory findings are included in Table III. Two of these patients have died. In neither was necropsy performed.

Group I

In these 5 patients the anatomical situation is an exaggeration of that in the tetralogy of Fallot or in the Eisenmenger complex. The aortic and mitral valves are contiguous through the ventricular septal defect, and the pulmonary trunk is normally situated beyond the crista.

Case 1. A boy was first seen in 1959 at the age of 8 months, having failed to thrive. He was acyanotic with a diffuse loud systolic murmur. There was a thrill at the aortic area. Pulmonary closure was not accentuated, and no apical mid-diastolic murmur was heard. Radiography showed cardiac enlargement and pulmonary plethora, while the electrocardiogram showed right axis deviation and probable left ventricular hypertrophy. Cardiac catheterization at that time showed a smallish left-to-right shunt at ventricular level with mild pulmonary valve stenosis (Table III). There was no arterial desaturation. Severe cardiac failure developed at the age of 3 years and 10 months. Further investigation at the age of 3½ years showed equalization of right ventricular and aortic systolic pressures, minimal, if any, left-to-right shunting, and evidence of mild pulmonary valve stenosis with moderate pulmonary arterial hypertension (Table III). The aortic catheter consistently entered the right ventricle but not the left. Direct left ventricular puncture was not attempted, despite a clinical diagnosis of left ventricular outflow obstruction, as it was felt that the boy's condition precluded any attempt at open-heart surgery.

Necropsy revealed a double outlet right ventricle. The pulmonary valve was normally placed, being separated from the aortic valve by a narrow cristall band (Fig. 1A, B). The aorta arose wholly from the right ventricle. The junction between the mitral and tricuspid valve rings formed part of the boundary of the ventricular septal defect, approximately 7 by 18 mm. in size, which provided the only outlet for the left ventricle

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TABLE I
CASES WITH NECROPSY

Case No. and sex	Age at death	Cyanosis	Pulmonary trunk	Ventricular septal defect	Other major defects
1 M	3 yr. 10 mth.	Nil	Normally situated	Subaortic	Bicuspid, moderately stenotic pulmonary valve
2 M	6 yr.	Moderate	Normally situated	Subaortic	Infundibular and pulmonary valve stenosis; secundum ASD
3 F	1 yr. 9 mth.	Mild	Normally situated	Subaortic	Secundum ASD; A-V canal with hypoplastic mitral valve component, small left ventricular cavity
4 F	2 wk.	Nil definite (jaundiced)	Normally situated	Subaortic	Secundum ASD; hypoplastic mitral valve and left ventricle
5 F	10 mth.	Progressive from infancy; ultimately deep	Normally situated	Subaortic	Severe infundibular stenosis
6 M	3 yr. 10 mth.	Mild to moderate*	Normally situated	Subaortic; tissue between mitral and aortic valves	Anomalous chordal attachment
7 M	9 yr.	Deep	Normally situated	Subaortic; tissue between mitral and aortic valves	Infundibular and pulmonary valve stenosis
8 M	4 mth.†	Mild	Transposed	Two defects, one subpulmonary	Anomalous chordal attachment
9 F	3½ wk.†	Minimal	Possibly transposed	Large subpulmonary defect	ASD; complete interruption of aortic arch
10 F	6 wk.†	Marked	Probably normally situated	Two defects, one largely occluded	Hypoplastic left lung

* Specimen from Pathology Department files. Notes record cyanosis during admissions with pneumonia and heart failure.

† Death precipitated by operation or by investigation.

TABLE II
PATIENTS SURVIVING OR WITHOUT NECROPSY FINDINGS

Case No. and sex	Age at, and year of, investigation	Cyanosis	Associated lesions	Course
11 M	17 mth., 1962	Mild	Left-to-right shunt at atrial level	Died, aged 3 yr., no necropsy
12 F	2 yr., 1962	Deep	Skeletal abnormalities; absence of hepatic IVC with azygos continuation; pulmonary valve stenosis	Satisfactory aorto-pulmonary anastomosis at 3 yr.
13 F	3 yr., 1963	Mild	—	Surgery not attempted
14 M	2 mth., 1963	Deep	Severe infundibular stenosis	Aorto-pulmonary anastomosis at 5 mth. poorly tolerated
15 M	3 mth., 1964	Mild	Atrial septal defect with large left-to-right shunt	Pulmonary artery banding without effect on haemodynamics
16 M	8 yr., 1965	Deep	Severe infundibular (subpulmonary) stenosis; apparent transposition of great vessels	Death following intracardiac surgery; necropsy not permitted

TABLE III
CARDIAC CATHETERIZATION FINDINGS

Case No.	Age	Oxygen saturations (%)							Pressures (mm. Hg)						
		Sup. vena cava	R. atrium	R. ventr.	Pulm. art.	L. atrium	Aorta	Ear-piece	R. atrium	R. ventr.	Pulm. art.	L. atrium	L. ventr.	Aorta	FA
1	8 mth.		55	71	70			97		55/0	25/10				
	3 yr.	59	53	65				93		90/0	50/30			90/55	
2	5 yr.	64	67	71		84*		80		80/0					
3	6 wk.	48	76	70	74			77		95/5	80/60				
4	10 dy.	34	91	86						45/5					
5	10 mth.	15		22		†				110/3					
9	3 wk.	40	76	78	87	93	87§	97	10 mean	90/20		17 mean		80/50	
									4 mean	90/5	82/42	13 mean			
11	17 mth.	59	78	75	75	98		87							
12	2 yr.	50	63	63		92		88		105/2					
13	3 yr.	69	65	92	90	§				100/0	100/50				90/58
14	2 mth.	26	26	45	96					90/10					
15	3 mth.	54	80	88		97			7 mean	75/10	—	12 mean	100/10		
	3 mth.	38	63	80	75	93‡	84			100/0	40/20¶			90/60	
16	8 yr.	62	66	80	85	100				100/10	16/6			(infundibulum 40/-4)	

* PV 98 per cent. † PV 96 per cent. ‡ PV 93 per cent. § Wedged PA 97 per cent. || Angiography confirmed obstruction to outflow to pulmonary artery (PA not entered by catheter). ¶ After banding pulmonary trunk.

FIG. 1.—(A) Right ventricular cavity viewed from anterior aspect showing apparently normal relationship of aorta and pulmonary trunk. (B) Right ventricular cavity viewed from below. Aortic valve visible behind crista, adjacent to tricuspid valve.

FIG. 1.—(C) Left ventricular cavity showing ventricular septal defect adjacent to mitral valve.

Diagram: m=mitral valve; v=ventricular septal defect; lsvc=left superior vena cava; p=left pulmonary artery.

(Fig. 1C). The aortic valve was adjacent to the atrio-ventricular valves, and the aortic cusp of the mitral valve was contiguous with the aortic valve through the septal defect. Both ventricles were dilated, particularly the right. The left ventricular wall measured 7 mm. and the right 8 to 10 mm. in thickness. The pulmonary valve was bicuspid and moderately stenotic.

Case 2. A boy was first seen during 1962, aged 5 years, with features suggestive of the tetralogy of Fallot with moderate cyanosis. The electrocardiogram showed right axis deviation and right ventricular hypertrophy. Investigation showed infundibular stenosis with a pulmonary trunk of moderate width. There was an interatrial communication (Table III). The aorta appeared conspicuously dextraposed with a left arch. Thoracotomy was performed with the object of total repair, but the aorta appeared to arise wholly from the right ventricle and this procedure was not attempted. Subsequently an anastomosis was made between the descending aorta and the left pulmonary artery. Following this, his condition became unsatisfactory with cardiac enlargement and evidence of failure. The anastomosis was narrowed at a further operation 6 weeks later, but heart failure persisted and he died 7 months after the initial shunt procedure. During this time clinical features suggested that pulmonary flow was not conspicuously increased.

Necropsy revealed a secundum atrial septal defect, and a prominent inferior vena caval valve. The aorta arose wholly from the right ventricle, being separated from the normally situated pulmonary valve by the crista. The aortic valve was contiguous with the mitral valve through a ventricular septal defect approximately 15 mm. in diameter that had as part of its boundary the junction of tricuspid and mitral rings. There was a short infundibular stenosis with a bicuspid pulmonary valve which was quite severely narrowed. There was a patent aorto-pulmonary anastomosis approximately 5 mm. in diameter. The left ventricle was dilated with a wall thickness of 10 mm., while the right ventricle was clearly hypertrophic with a thickness of 12 mm.

Case 3. A female infant was seen first in April 1962, when aged 1 month, with cardiac failure and signs of pulmonary hypertension with considerable cardiac enlargement and pulmonary plethora. There were also multiple skeletal abnormalities, with hemivertebrae and fused ribs. The electrocardiogram showed marked right ventricular hypertrophy with an apparent QR complex in the right chest leads. Investigations showed substantial left-to-right shunt at atrial level with right ventricular hypertension (Table III). She thrived poorly and had persistent dyspnoea. Death occurred at 21 months apparently from inhalation of vomitus associated with a fit.

Necropsy revealed dilatation of the right atrium with a large secundum atrial septal defect and a primum atrial septal defect. Both main trunks arose from the right ventricle, the pulmonary artery being normally situated anterior to the crista. The aortic ring was contiguous with a common atrio-ventricular valve whose mitral component was very hypoplastic. A ventricular septal

defect provided egress from the left ventricle. This defect was roofed by the fused anterior and posterior leaflets of the common atrio-ventricular valve. The right ventricle was dilated and hypertrophic, while the left ventricle was relatively small with the cavity decreased in size by septal bulging.

Case 4. A female infant was admitted in March 1964 at the age of 5 days with cardiac failure. Her condition progressively deteriorated, and she died at the age of 2 weeks. The radiograph showed marked cardiac enlargement and apparent pulmonary venous engorgement. The electrocardiogram showed right ventricular dominance with a small "q" wave in the right-sided chest leads. Cardiac catheterization demonstrated a left-to-right shunt at atrial level with right ventricular hypertension (Table III). Both aorta and pulmonary artery filled from an anteriorly-placed ventricle into which radio-opaque dye was injected for selective angiography.

Necropsy revealed a secundum atrial septal defect. The pulmonary artery was normally placed. The aorta also arose from the right ventricle, its root being separated from the pulmonary valve by cristal tissue. Mitral and tricuspid valves were contiguous through a small ventricular septal defect which provided the only outlet for the left ventricle. The aortic ring was in virtual continuity with the mitral ring through the defect. The mitral valve was narrow and the left ventricle somewhat hypoplastic.

Case 5. A female infant was seen first in February 1964 at 4 weeks of age. There was no cyanosis. A loud præcordial systolic murmur radiated to the aortic area. There was a probable right aortic arch and the lung fields were somewhat dry. The electrocardiogram suggested excess left ventricular activity. During subsequent months cyanosis became progressively more marked and disability more severe, but the murmur did not change. At 10 months the ear-piece saturation was 56 per cent at rest. Selective right ventricular angiography showed severe infundibular stenosis with a ventricular septal defect, the aorta arising wholly from the right ventricle. Necropsy confirmed these findings together with severe pulmonary valve stenosis. The aortic and mitral valves were contiguous as in the tetralogy of Fallot, the aortic root simply being dextraposed fully across to the right ventricular side of the septum.

Group II

This group comprises 2 patients in whom the pulmonary trunk was normally situated with a ventricular septal defect proximal to the crista and close to the atrio-ventricular valves. In contrast to Group I the aortic and mitral valves were separated by an obvious band of tissue.

Case 6. This boy died in 1954 at the age of 3 years of congestive cardiac failure, having previously suffered from recurrent lower respiratory tract infection associ-

FIG. 2.—(Case 6). Right ventricular cavity viewed from anterior aspect. Aortic valve separated from septal defect and atrio-ventricular valves by band of muscle.

ated with known congenital heart disease. Necropsy revealed double outlet right ventricle.

The tricuspid ring was normal but the lateral cusp of the tricuspid valve was small. The anterior and septal cusps fused medially and formed the posterior margin of a ventricular septal defect, the superior margin of which was formed by the aortic cusp of the mitral valve (Fig. 2). Both aorta and pulmonary trunk arose from the somewhat thickened right ventricle, the pulmonary trunk lying normally beyond a hypertrophied crista. The aortic root was separated from the aortic cusp of the mitral valve by a band of muscle approximately 10 mm. in width. The endocardium overlying this muscle band was thickened and fibro-elastic. The left ventricle was hypertrophied and dilated, the only egress being via the ventricular septal defect. The mitral valve was somewhat thickened with short chordæ.

Case 7. A boy aged 9 years, previously investigated and thought to have a severe form of the tetralogy of Fallot, died suddenly at home. Necropsy confirmed the presence of severe localized infundibular stenosis with calcification. There was a narrow bicuspid pulmonary valve and the pulmonary trunk was small. The aorta arose wholly from the right ventricle proximal to the crista, its root being separated by a band of muscle approximately 10 mm. wide from the aortic cusp of the mitral valve, where that cusp met the tricuspid valve in the edge of the ventricular septal defect.

Diagram: a=aorta; p=pulmonary trunk; mb=muscle band; t=tricuspid valve; v=ventricular septal defect.

Group III

In this patient there appeared to be transposition of the great vessels complicating the double outlet right ventricle.

Case 8. A male child presented with cardiac failure at the age of 4 months. He had a loud præcordial systolic murmur with a thrill and was mildly cyanosed. Radiography showed considerable cardiac enlargement with pulmonary plethora, while the electrocardiogram showed right axis deviation, biventricular hypertrophy, and right atrial hypertrophy. Death resulted from cerebral oedema following a venous angiocardigram (Case 11, Venables and Hiller, 1963).

Necropsy showed a dilated right atrium with multiple small perforations in the floor of the fossa ovalis. There was a double outlet right ventricle. The aortic root was transposed, lying beyond the crista, while the pulmonary valve was situated more posteriorly, lying close to the tricuspid valve, pulmonary valve and mitral valve being contiguous through a ventricular septal defect (Fig. 3A, B). Across the right ventricular aspect of this ventricular septal defect ran a thick muscular band which extended from the apex of the right ventricle to just below and lateral to the pulmonary valve dividing the defect into two substantial components, the medial of which was that related to the pulmonary trunk, as described above. The lateral defect was partly occluded on its left ventricular aspect by anomalous attachment of the mitral valve to the related part of the septum. The left ventricle was dilated and the right ventricle moderately hypertrophied.

Group IV

In these two cases the anatomical situation is more difficult to define. In one, there was a large subpulmonary ventricular septal defect. In the

FIG. 3.—(Case 8). Right ventricular cavity viewed from anterior aspect.
 (A) Right ventricle partly unroofed showing relationship of aorta and pulmonary trunk.
 (B) Right ventricle wholly unroofed showing two components of ventricular septal defect. Pulmonary valve is continuous with mitral valve through medial defect adjacent to tricuspid valve.

Diagram: a = aorta; p = pulmonary trunk; t = tricuspid valve; v = ventricular septal defect; mb = muscle band across defect.

other there were 2 ventricular septal defects, of which the major functional defect lay in the muscular septum well away from either aorta or pulmonary trunk.

Case 9. A female infant developed cardiac failure at the age of 1 week. There were no significant murmurs. The ear-piece saturation was 91 per cent at rest in air, rising to 97 per cent in oxygen. The radiograph showed cardiac enlargement and pulmonary plethora. The electrocardiogram was within normal limits for age.

Investigation suggested the presence of both an atrial septal defect and a ventricular septal defect, with right ventricular systolic pressures of systemic order. The catheter passed from the pulmonary trunk to the descending aorta where the saturation equalled that in the pulmonary artery (Table II). Death occurred at thoracotomy during the fourth week of life when an attempt was being made to reduce the left-to-right shunt.

Necropsy revealed an atrial septal defect of the foramen ovale type, a double outlet right ventricle, and complete interruption of the aortic arch beyond the left subclavian artery. The descending aorta was continuous with the pulmonary trunk. Fig. 4 shows the rela-

tionship of tricuspid valve, aortic root, pulmonary trunk, and ventricular septal defect as viewed from the right ventricle. There was a large subpulmonary ventricular septal defect. Through this defect the tricuspid valve tissue was contiguous with the mitral valve. There was a band of muscle between the mitral ring and the pulmonary ring and also between tricuspid and aortic rings.

Case 10. A female infant presented in January 1961 at the age of 2 weeks with cyanosis and dyspnoea since birth, and apparent cardiac failure. Ear-piece saturation was 72 per cent. There was a faint præcordial systolic murmur. Radiography showed marked cardiac enlargement and moderate pulmonary plethora, while the electrocardiogram showed right ventricular preponderance associated with dominant "s" waves in leads II, III, and aVF. A venous angiogram, misinterpreted at the time, showed the aorta and pulmonary trunk filling from the right ventricle without left ventricular filling, the aorta being placed anteriorly and somewhat medially.

The baby died during thoracotomy at the age of 6 weeks. Necropsy revealed a rather anteriorly placed aorta arising from the right ventricle. The aorta was separated from the tricuspid valve by a wide band of

FIG. 4.—(Case 9). Right ventricular cavity viewed from anterior aspect. Large subpulmonary ventricular septal defect.

muscle which was a continuation of the crista supraventricularis. The pulmonary trunk also arose from the right ventricle, its valve being separated from the aortic root and from the right ventricular inflow by the crista (Fig. 5A). The right and particularly the left ventricle were both dilated. The only effective egress from the left ventricle was via a septal defect approximately 10×7 mm. situated in the muscular septum well away from the tricuspid valve below the crista supraventricularis (Fig. 5B). An additional defect in the region of the atrio-ventricular valves when viewed from the left ventricle was virtually sealed by an adherent somewhat anomalous mitral valve. The right ventricular opening of the non-functional septal defect was small and some distance below the pulmonary valve ring. The left lung was hypoplastic with a single pulmonary vein.

Group V

Details of the surviving cases, 11 to 16 inclusive, are summarized in Table II.

DISCUSSION

Anatomical Findings. Cases of double outlet right ventricle have been classified in two ways, depending first on the presence of significant pulmonary stenosis (Witham, 1957; Neufeld *et al.*,

Diagram: a=aorta; p=pulmonary trunk; t=tricuspid valve; v=ventricular septal defect.

1961a, b, 1962; Morgan *et al.*, 1962), and second on the relation between the aortic and mitral valves (Neufeld *et al.*, 1961b, 1962).

Neufeld *et al.* (1961b) described two subgroups of double outlet right ventricle without pulmonary stenosis. In 3 of 5 specimens, the aortic leaflet of the mitral valve was long and ran through the roof of the ventricular septal defect to become continuous with the aortic valve. In the other 2 specimens, the aortic and mitral valves were separated by a band of muscle described as part of the crista supraventricularis. The ventricular septal defects in these individuals were situated proximal to the crista, adjacent to the atrio-ventricular valves. In the later, different, series of Neufeld *et al.* (1962), the aortic valve was continuous with the mitral valve in only 1 of the 6 specimens without pulmonary stenosis and with a septal defect in this position. Continuity of aortic and mitral valves was present in only 1 of 6 cases with double outlet right ventricle, complicated by pulmonary stenosis (Neufeld *et al.*, 1961a). In the others the aortic valve was noted to be above the horizontal limb of the crista supraventricularis which separated the aortic valve from the ventricular septal defect and from the atrio-ventricular valve tissue.

Van Mierop and Wiglesworth (1963) also referred to these two anatomical types in discussing the embryology of double outlet right ventricle. Whereas Neufeld *et al.* (1962) merely stated that "discontinuity of the mitral and aortic valve tissue is the usual finding" in double outlet right ventricle without pulmonary stenosis, Van Mierop and

FIG. 5.—(Case 10).
 (A) Right ventricular cavity viewed from anterior aspect. (B) Left ventricular cavity showing major (functional) ventricular septal defect.

Diagram: a=aorta; p=pulmonary trunk; t=tricuspid valve; v=virtually non-functioning ventricular septal defect; v¹= major ventricular septal defect.

Diagram; mv=mitral valve; v=virtually non-functioning ventricular septal defect; v¹=major ventricular septal defect.

Wiglesworth (1963) regarded the type with contiguous aortic and mitral rings as merely an extreme form of the Eisenmenger complex, or of the tetralogy of Fallot if there were significant pulmonary stenosis, embryologically different from the type in which aortic and mitral valves are clearly separated by muscle. These authors referred to this separating tissue as persistence of the bulboventricular flange. Despite their observations they admitted

the functional identity of the two types, which can only be differentiated by direct observation of the defect and the aortic ring at operation or necropsy.

In the present series there were 7 hearts in which the pulmonary trunk was clearly situated in normal relation to the crista and to the other valves (Cases 1 to 7). Three exhibited severe outflow tract stenosis of mixed infundibular and valvular type, while one had a moderately stenotic bicuspid

pulmonary valve. In 4 specimens there was continuity between the mitral and aortic valves, and in one with a persistent common atrio-ventricular canal there was continuity between the canal valve and the aortic valve. In the remaining 2 specimens (Cases 6 and 7), the aortic valve was separated from the mitral valve by a flat band of muscle approximately 10 mm. wide. In one of these there was severe pulmonary stenosis. Thus all but 2 of these specimens appear to be examples of severe aortic dextroposition of the Eisenmenger and Fallot types in which the aorta arises wholly from the right ventricle. In all 7, the septal defect was proximal to the crista.

In 3 hearts the anatomy was more complex. In Case 8 the great vessels appeared certainly to be transposed with the aorta lying beyond the crista, the pulmonary trunk having an essentially posterolateral relationship to the aorta, with its valve close to the atrio-ventricular valves. One component of the ventricular septal defect was subpulmonary in position and there was a close relation between pulmonary and mitral valves (Fig. 3A and B). Grant (1962) emphasized the presence in transposition of the great vessels of an intimate relation between the pulmonary and mitral rings in contrast to the normal close relation of aortic and mitral rings. By this criterion Case 8 exhibits transposition independent of any consideration or identification of exact right ventricular architecture. In Case 9 (Fig. 4) the aorta arose from a muscular tunnel medial to the pulmonary trunk and the vessels were less definitely transposed. The septal defect was sub-pulmonary (Fig. 4) but without contiguity of pulmonary and mitral valves. Subpulmonary septal defects occur without transposition, and Case 9 probably fits best into this category. Neufeld *et al.* (1962) described 3 cases of double outlet right ventricle with ventricular septal defects lying above the crista supraventricularis in a subpulmonary position. In one the defect extended below the aortic valve which was continuous with the aortic cusp of the mitral valve. In the others absence of continuity between aortic and mitral valves was mentioned without reference to possible relationship between pulmonary and mitral valves. There is a striking similarity between the right ventricular aspect of their Case 8 as illustrated and Fig. 4 of this report. Clear-cut transposition was stated to be present in the case with persistent common atrio-ventricular canal and pulmonary stenosis described by Neufeld *et al.* (1961a).

In Case 10 of this report the main trunks were probably not transposed. There were two ventricular septal defects of which one was probably of little functional significance. This smaller defect

passed from below the aortic cusp of the mitral valve on the left ventricular aspect of the septum to a point a short distance below the pulmonary valve. The major defect was lower in the muscular septum. As in Case 9, there was no direct relation between the mitral valve and either semilunar valve. This situation was in fact shared by the two cases (Cases 6 and 7) of Group II. Grant (1962) acknowledged the occurrence in some examples of double outlet right ventricle of this departure from his scheme of valve ring relationships.

In the specimens described in this report, there were three categories of valve relationships. An intimate relation between aortic and mitral valves, as in the normal heart, was most common, but in one specimen there was a close relation between pulmonary and mitral rings, as in complete transposition. In 4 specimens there was no intimate relation between the mitral valve and either the aortic or pulmonary valves. These patterns provide an important method of classification of the anatomy of the condition.

Hæmodynamic Disturbances. The presence of a subpulmonary ventricular septal defect has physiological implications that are independent of whether or not the great vessels appear to be anatomically transposed. In the more usual form of double outlet right ventricle where the aorta is closely or relatively closely related to the septal defect, the hæmodynamic situation is frequently similar to that of large ventricular septal defect. Neufeld *et al.* (1961b) pointed out that the impression of direct continuity of the aorta with the left ventricle, given by opening these specimens from left ventricle to aorta, was an artefact, but despite this the aorta receives left ventricular blood. Unless there is significant pulmonary stenosis or pulmonary vascular disease there appears in many cases to be little entry of systemic venous blood from the right ventricle into the aorta, and arterial saturations are often relatively normal. There is obligatory right ventricular hypertension with substantial left-to-right shunt. On the other hand if the septal defect is subpulmonary, the left ventricle ejects directly into the pulmonary artery and the aorta receives largely systemic venous blood, so that substantial cyanosis is combined with considerable increase in pulmonary flow and very high oxygen saturations in the pulmonary artery. The situation is hæmodynamically like that of many cases of complete transposition of the great vessels. When the pulmonary trunk is normally situated and the septal defect is subpulmonary, the abnormality is held by Neufeld *et al.* (1962) and by Van Mierop and Wigglesworth (1963) to represent the Taussig-

Bing syndrome in which the pulmonary trunk is more commonly described as overriding the ventricular septum above the defect.

Cases 8 and 9 of the present report exhibit subpulmonary defects. Case 9 (Fig. 4) more closely resembles the Taussig-Bing syndrome than Case 8 (Fig. 3) in which the situation appears to be actually more closely related to transposition with ventricular septal defect and overriding pulmonary artery. In both cases, however, the pulmonary artery is arising wholly from the right ventricle and not overriding. It is of interest that neither of these patients in fact had deep cyanosis. In Case 9 this appears to have been due to a large left-to-right shunt at atrial level resulting in a high oxygen saturation of blood entering the right ventricle from the right atrium. Case 8 was studied only by venous angiography so that the haemodynamic situation was not documented, but it is possible that the second ventricular septal defect or the multiple small atrial defects may have influenced the aortic saturation.

The only patient in the series with deep cyanosis without severe pulmonary stenosis was Case 10. In this specimen the functional ventricular septal defect was a considerable distance from both aorta and pulmonary artery, so that it is difficult to account for the aortic saturation in terms of specific direction of the stream of left ventricular blood.

As already stated, in the common forms of double outlet right ventricle the left ventricle ejects blood directly into the aorta as in simple ventricular septal defect with normal aortic root. Complication by severe pulmonary stenosis or pulmonary vascular obstruction results in diversion of part of the systemic venous return from the right ventricle to the aorta with consequent arterial desaturation and cyanosis. In double outlet right ventricle, however, the left ventricular output has all to pass through the septal defect, which occupies a relationship to the ejected blood comparable to the obstructive tissue in the syndrome of ventricular septal defect with subaortic stenosis situated at the lower level of the defect. The septal defect may be obstructive both with normal or reduced left ventricular output and with the increased output present when pulmonary flow is increased naturally or as the result of aorto-pulmonary anastomosis. Mirowski, Mehrizi, and Taussig (1963) emphasized the contribution of obstructive left ventricular loading to the electrocardiographic picture of double outlet right ventricle with significant pulmonary stenosis, while Taussig (1960) has commented on the poor tolerance of these patients for shunt procedures. Both of these phenomena are likely to be due to interference with left ventricular out-

flow. Shunts were poorly tolerated in Case 2 and in the surviving Case 14 in spite of lack of clinical evidence of excessive pulmonary flow. In Case 1 the clinical course could not be explained in terms of the amount of shunt and the degree of pulmonary stenosis demonstrated, and aortic stenosis was actually thought to be the additional responsible factor.

The relationship of the septal defect to the left ventricular outflow stream is probably also responsible at times for the murmurs observed in some patients with this condition. Murmurs suggestive of aortic stenosis were described in cases of double outlet right ventricle without pulmonary stenosis by Morgan *et al.* (1962) who commented on "aortic outlet narrowing" in three patients with such murmurs. In Case 1 the murmur suggested the presence of aortic stenosis, but at necropsy there was no aortic valve abnormality and no "aortic outlet narrowing", the aortic valve being contiguous with the mitral valve. The septal defect appeared to have provided obstruction to outflow from the left ventricle and was probably the site of origin of the murmur. In Case 5 a loud præcordial systolic murmur, which persisted when pulmonary flow appeared to have become very small indeed, again seems likely to have been generated at the septal defect.

Substantial left-to-right shunt was present at atrial level in 5 of the patients who were catheterized (Table III). In 3 the mean left atrial pressure exceeded the mean right atrial pressure by 5 to 9 mm. mercury. The 3 who died had large atrial septal defects. In Case 15 the persistence of the atrial shunt after effective pulmonary artery banding and the findings at subsequent angiographic studies also indicated that there was a true atrial septal defect rather than a dilated foramen ovale, permitting left-to-right shunt consequent on dilatation of the left atrium secondary to increased pulmonary flow, as described by Rudolph *et al.* (1958). In two of those who died there was evidence of obstruction to left ventricular filling that would have potentiated the shunt.

Electrocardiograms. Neufeld *et al.* (1961b) described an electrocardiographic pattern similar to that of persistent common atrio-ventricular canal in 7 of 8 patients without pulmonary stenosis. In the series of Neufeld *et al.* (1962) this pattern occurred in similar patients whose ventricular septal defects lay below the crista. This pattern was not discussed in the cases with pulmonary stenosis of Neufeld *et al.* (1961a) and of Mirowski *et al.* (1963). The findings in the present series showed no constant pattern but do not negate the possible value

of such an electrocardiogram as a diagnostic clue in apparent ventricular septal defect with pulmonary hypertension.

Diagnosis. The possibility of double outlet right ventricle arises in all patients with apparent ventricular septal defect with right ventricular hypertension of systemic level, and in patients who appear to have the tetralogy of Fallot with extreme dextroposition of the aorta. The special surgical problems of the condition (Kirklin, Harp, and McGoon, 1964) make it important that the anatomical situation be recognized by appropriate investigatory procedures, and that possible left ventricular outflow obstruction be assessed whenever indicated. The diagnosis may be suggested by the anterior position of the aortic root, by vertical orientation of that structure, as suggested by Morgan *et al.* (1962), or by consistent passage of an aortic catheter into the right but not the left ventricle. The diagnosis must, however, be confirmed by appropriate biplane angiography demonstrating the relationship of the great vessels to the right ventricle, remembering the dilution effects which may result from selective left ventricular ejection to one or other main trunk (Neufeld *et al.*, 1962). In one case in the present series selective left ventricular angiography performed via the foramen ovale demonstrated ejection of the left ventricular contents via the ventricular septal defect to the right ventricle from which both main trunks arose. Investigation will reveal double outlet right ventricle, possibly complicated by transposition, in some infants presenting arterial desaturation with increased pulmonary flow but with no clear clinical indication of the nature of the underlying lesion.

SUMMARY

Sixteen cases of double outlet right ventricle seen at the Royal Children's Hospital, Melbourne, are recorded. The diagnosis was based on necropsy in 10 cases and on investigatory findings alone in 6 cases.

The patterns of the anatomy of the condition are illustrated by the 10 necropsy specimens. In 5 specimens there was a normal relation between mitral and aortic valve tissue through the ventricular septal defect that provided the only egress from the left ventricle. In one specimen there was a similar close relation between pulmonary and mitral valves, as in transposition of the great vessels. In the remaining 4 specimens there was no contiguity between either the aortic or pulmonary valves and the atrio-ventricular valve tissue. These relationships appear to provide an important method of classi-

fying specimens of double outlet right ventricle in addition to the two other methods employed, related to the presence or absence of pulmonary stenosis and to the relationship of the ventricular septal defect to the aorta or pulmonary artery. The details of the anatomy have considerable relevance to the possibility of intracardiac repair, but final assessment can only be made by direct inspection of the interior of the heart.

The patterns of physiological disturbance in double outlet right ventricle are also discussed in relation to the material reported. The site and size of the ventricular septal defect are of considerable importance. Selective direction of left ventricular outflow to one or other great vessel greatly influences systemic arterial saturation, while obstruction to left ventricular outflow by the septal defect affects left ventricular loading.

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