THE HÆMODYNAMICS OF COMPLETE TRANSPOSITION OF THE GREAT VESSELS

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In the normal circulation blood goes around the body in one circuit. Two resistances are encountered by the blood in this circuit, the systemic and the pulmonary vascular resistances. In complete transposition of the great vessels, blood goes round in two separate circuits, namely the systemic and pulmonary circuits. Owing to the separate nature of the two circuits, any shunt that occurs between them has to be bidirectional and exactly equal in both directions. Theoretically if a large communication, such as a ventricular septal defect, exists between the two circuits, a shunt occurs from the circuit with the higher pressure to that with the lower pressure only until the pressures in the two ventricles equalize. The fact that the pulmonary and systemic resistances are often different in complete transposition of the great vessels will not result in itself in the exchange of blood between the two circuits, because even if it explains the direction of the shunt from the circuit with the higher resistance to that with the lower resistance, it will not explain the occurrence of an equal shunt in the reverse direction, that is from the circuit with the lower resistance to that with the higher.

Several views have been expressed about the mechanism of blood shunting in transposition of the great vessels (Astley and Parsons, 1952; Wood, 1956; Kjellberg *et al.*, 1959; Taussig, 1960). These authors thought that the volume and the direction of the shunt in transposition of the great vessels was probably determined by the total resistances in the two vascular systems. The reason why some of the deeply cyanosed patients have evidence of increased systemic blood flow (Campbell and Suzman, 1951; Astley and Parsons, 1952; Cleland *et al.*, 1957; Kjellberg *et al.*, 1959; Noonan *et al.*, 1960; D. C. Deuchar, 1962, personal communication) has never been explained.

The object of this paper is to discuss the hæmodynamics of complete transposition of the great vessels. The belief that cardiac catheterization contributes little to the diagnosis of this condition (Campbell *et al.*, 1949; Blalock and Hanlon, 1950; Lillehei and Varco, 1953; Keith, Rowe, and Vlad, 1958) has probably led to the scarcity of the data in any one hospital. For this reason the majority of the patients studied hæmodynamically in this paper have been obtained from published reports, particularly from the excellent paper by Noonan *et al.* (1960).

SUBJECTS

There were 37 patients with complete transposition of the great vessels, for each of whom hæmodynamic data were available. Twenty-six patients have been collected from earlier papers (Campbell *et al.*, 1949; Lenkei, Swan, and DuShane, 1959; Noonan *et al.*, 1960). The other 11 patients were studied at Guy's Hospital and at Southampton Chest Hospital. In these 11 patients the diagnosis was confirmed by cardiac catheterization in 2, angiography in 2, cardiac catheterization and operation in 1, angiography and operation in 1, and by necropsy in the remaining 5.

For purposes of discussion these patients have been divided into two groups. Group 1 (Table IA and IB): 9 patients with a closed ventricular septum; Group 2: patients with a ventricular septal defect, subdivided into three groups, 2a, 2b, and 2c; Group 2a (Table IIA and IIB): 13 patients with the pulmonary blood flow

TABLE IA

PERCENTAGE BLOOD OXYGEN SATURATIONS OF NINE PATIENTS WITH CLOSED VENTRICULAR SEPTUM

No.	Reference	Percentage blood oxygen saturations						
		S.V.C.	R.A.	R.V.	Aorta	L.A.	L.V.	P.A.
1	Noonan 1.1	29	60	52	52	94	94	—
2	Noonan 1.2	44	61	65	66	93	94	—
3	Noonan 1.4	19	26	36	36	*88	94	—
4	Noonan 1'11	47	61	66	62	91	91	—
5	Linkei (Case 2) ..	59	77	69	71	90	90	—
6	Noonan 1.5	24	26	35	37	*86	92	—
7	Noonan 1.8	3.3	15	29	25	*99	85	—
8	Noonan 1.12	49	57	65	65	84	85	—
9	V.S.	56	72	70	70	*94.6	81	—

* Left ventricular sample used to assess right-to-left shunt.

S.V.C.—Superior vena cava; R.A.—right atrium; R.V.—right ventricle; L.A.—left atrium; L.V.—left ventricle
P.A.—pulmonary artery.

TABLE IB

PRESSURES AND BLOOD FLOWS OF NINE PATIENTS WITH CLOSED VENTRICULAR SEPTUM

No.	Blood pressures (mm. Hg)						Blood flows (l./min./m. ²)		
	Aorta	P.A.	R.V.	L.V.	R.A.	L.A.	Systemic	Pulmonary	Effective
1	75/40	—	75/—	75/5	5.0	11.0	5.0	—	1.2
2	65/40	—	75/5	65/12	5.0	22.0	5.0	—	2.1
3	80/42	—	80/12	50/4	4.0	5.0	7.0	—	1.4
4	110/65	—	93/5	69/19	5.5	6.5	3.2	8.0	1.1
5	88/67	—	86/6	37/0	5.0	17.7	7.0	—	2.15
6	90/50	—	95/12	35/5	8.0	8.0	8.2	—	1.7
7	70/40	—	77/4	85/2	10.0	7.0	3.6	6.4	0.9
8	105/75	—	100/—	90/12	6.0	6.0	4.3	4.0	1.1
9	80/52	—	82/—2	50/—1	2.0	2.0	3.2	3.8	1.1

exceeding the systemic blood flow; Group 2b (Table IIIA and IIIB): 8 patients with the systemic blood flow exceeding the pulmonary blood flow; Group 2c (Table IVA and IVB): 7 patients from whom only some of the necessary data were obtained.

A left-to-right shunt at atrial level was considered to be present when there was an increase of 8 per cent or more in the oxygen saturation at this level. A right-to-left shunt was considered to be present when the sample of blood from the left side of the heart had an oxygen saturation of 5 per cent or more, less than the pulmonary venous sample or less than 90 per cent if a pulmonary venous sample was not obtained.

RESULTS

In complete transposition of the great vessels, some method of oxygenating the blood in the systemic circuit must exist. This is achieved by exchanging part of the mixed venous blood in the systemic circuit with an equal volume of pulmonary venous blood from the pulmonary circuit, through the associated defects. This shunt, therefore, is the effective pulmonary blood flow, since it is the volume of mixed venous blood which, after returning to the right atrium, eventually reaches the pulmonary capillaries (Campbell *et al.*, 1949). The actual pulmonary blood flow, on the other hand, is the volume of blood flowing through the pulmonary artery per minute.

Flows. In the whole group of 37 patients, the effective pulmonary blood flow, i.e. the shunt, varied from 0.8–2.8 l./min./m.² with an average of 1.62 l./min./m.². In the group of 9 patients with a closed ventricular septum, the effective pulmonary blood flow averaged 1.4 l./min./m.². In the

TABLE IIA

BLOOD OXYGEN SATURATIONS OF 13 PATIENTS WITH VENTRICULAR SEPTAL DEFECTS AND PULMONARY BLOOD FLOWS EXCEEDING SYSTEMIC BLOOD FLOWS

No.	Reference	Percentage blood oxygen saturations						
		S.V.C.	R.A.	R.V.	Aorta	L.A.	L.V.	P.A.
10	Noonan 2C.2...	50	72	81	74	95	—	87
11	Noonan 2C.3...	44	66	—	82	94	93	90
12	Noonan 2C.9...	45	56	62	69	—	96	87
13	Noonan 2C.11	22	36	52	63	89	91	82
14	Noonan 2C.4...	41	47	70	80	P.V. 86	95	87
15	Noonan 2C.6...	37	44	66	61	98	96	91
16	Noonan 2B.6...	54	59	64	72	97	95	91
17	Noonan 2B.8...	14	17	35	28	94	94	84
18	Noonan 2A.9...	56	60	67	72	100	92	89
19	Noonan 2C.10	—	44	66	69	99	77	86
20	A.B. ...	34	30	55	46	—	—	88
21	E.T. ...	60	53	75	70	—	—	82
22	H.K. ...	53	50	61	70	—	—	84

P.V.—Pulmonary vein; other abbreviations as in Table IA.

TABLE IIB

PRESSURES AND FLOWS OF 13 PATIENTS WITH VENTRICULAR SEPTAL DEFECTS AND PULMONARY BLOOD FLOWS EXCEEDING SYSTEMIC BLOOD FLOWS

No.	Blood pressures (mm. Hg)						Blood flows (l./min./m. ²)		
	Aorta	P.A.	R.V.	L.V.	R.A.	L.A.	Systemic	Pulmonary	Effective
10	87/50	44/12	90/4	—	4.0	8.0	5.3	16.4	2.8
11	95/65	55/25	105/—	110/8	6.0	6.0	4.4	18.0	2.6
12	68/38	43/16	68/5	65/—	1.5	P.C.10.0	5.1	14.9	2.1
13	54/28	44/12	90/3	90/—	3.0	12.0	3.6	14.6	2.1
14	80/50	60/15	75/5	75/5	3.0	9.0	3.1	12.4	2.1
15	67/42	45/15	73/0.5	75/—	1.0	2.5	3.6	15.0	1.4
16	92/56	92/50	94/0	92/2	—3	0.0	5.0	8.2	2.2
17	80/55	100/60	100/12	100/10	4.0	5.0	4.7	5.0	0.8
18	90/43	m 13	85/5	100/5	4.0	6.0	4.0	6.0	1.5
19	85/54	64/20	82/6	88/10	0.0	8.0	4.0	13.0	1.9
20	68/34	67/28	72/—1	—	5.0	P.C.11.0	8.9	15.0	1.6
21	—	m 88	—	—	10.0	—	4.3	9.2	2.0
22	100/6	94/54	93/0	100/0	2.0	—	4.0	6.7	1.8

P.C.—Pulmonary capillary; m—mean; other abbreviations as in Table IA.

group of 13 patients with a ventricular septal defect and the pulmonary blood flow exceeding the systemic blood flow, the shunt averaged 1.93 l./min./m.². In the group of 8 patients with a ventricular septal defect and the systemic blood flow exceeding the pulmonary blood flow, the shunt averaged 1.5 l./min./m.². The pulmonary and systemic flows, however, varied within a much wider range. The pulmonary blood flow in the group of 27 patients in whom this measurement was available ranged from 1.7–18.0 l./min./m.², while in the whole group of 37 patients, the systemic blood flow ranged from 3.2–18.0 l./min./m.².

Direction of the Shunt. Group 1 (Table IA and IB) (patients with a closed ventricular septum). Patients 1, 2, 3, 4, 5, and 6 only had a left-to-right shunt at atrial level, the right-to-left shunt presumably being at pulmonary artery level (patent ductus arteriosus or bronchial collaterals). Patients

TABLE IIIA

BLOOD OXYGEN SATURATIONS OF 8 PATIENTS WITH VENTRICULAR SEPTAL DEFECTS AND SYSTEMIC BLOOD FLOWS EXCEEDING PULMONARY BLOOD FLOWS

No.	Reference	Percentage blood oxygen saturations						
		S.V.C.	R.A.	R.V.	Aorta	L.A.	L.V.	P.A.
23	Noonan 2B.4	25	24	26	32	91	85	—
24	Noonan 2A.3	32	32	33	42	82	75	—
25	Noonan 2A.4	49	56	63	66	88	78	—
26	Noonan 2B.1	58	61	—	70	92	87	85
						P.V. 97		
27	Noonan 2B.2	9	8	8	13	81	84	81
28	Noonan 2A.1	45	44	50	56	68	69	—
29	Noonan 2A.5	42	36	44	50	56	54	—
30	P.M.	64	63	—	74	83	83	—

P.V.—Pulmonary vein; other abbreviations as in Table IA.

TABLE IIIB

BLOOD PRESSURES AND BLOOD FLOWS IN 8 PATIENTS WITH VENTRICULAR SEPTAL DEFECTS AND SYSTEMIC BLOOD FLOWS EXCEEDING PULMONARY BLOOD FLOWS

No.	Blood pressures (mm. Hg)						Blood flows (l./min./m. ²)		
	Aorta	P.A.	R.V.	L.V.	R.A.	L.A.	Systemic	Pulmonary	Effective
23	95/43	100/58	100/10	95/8	4.5	3.5	12.0	4.5	1.4
24	94/71	P.S.	107/11	94/9	5.0	4.5	7.8	3.0-4.9	1.1
25	109/64	P.S.	95/5	95/3	4.0	3.0	4.0	3.0	1.4
26	98/63	103/63	—	100/—	5.0	5.0	6.0	5.3	1.8
27	95/45	90/50	90/5	93/2	5.0	3.0	18.0	6.0	1.0
28	105/65	P.S.	95/0	100/7	7.0	6.5	5.5	2.2	1.1
29	95/50	P.S.	95/8	—	7.5	5.5	5.3	1.7	1.2
30	75/5	P.S.	—	90/0	0.0	0.0	8.3	7.6	2.75

P.S.—Pulmonary stenosis; other abbreviations as in Table IA.

7, 8, and 9 had a bidirectional shunt at atrial level. None of the patients showed a unidirectional right-to-left shunt at atrial level.

Group 2a (Table IIA and IIB). (Patients with a ventricular septal defect with the pulmonary blood flow exceeding the systemic flow (i.e. patients with mild pulmonary stenosis or low pulmonary vascular resistance).)

The left-to-right shunt: In Patients 10, 11, 12, 13, 14, 15, 16, 17, 18, and 19 where blood samples were obtained from the left side of the heart, a left-to-right shunt at atrial level was found in the first 4 patients and further increase in the blood O₂ saturation was noted in the right ventricle. In Patients 14, 15, 16, 17, and 18 with no form of blood shunting at atrial level, and in Patient 19 where a superior vena cava sample was not obtained, a left-to-right shunt at ventricular level was present.

The right-to-left shunt: A left ventricular sample was obtained in Patients 11, 12, 13, 14, 15, 16, 17, 18, and 19. Whereas only the last 2 patients showed desaturation of the left ventricular sample, this sample was fully saturated in the remaining 7 patients (11, 12, 13, 14, 15, 16, and 17). In these 7 patients the right-to-left shunt could be either direct from the right ventricle to the pulmonary artery through the ventricular septal defect or from the aorta to the pulmonary artery through anastomotic collaterals or a patent ductus arteriosus. The last assumption is only possible if a pressure gradient exists between the aorta and the pulmonary artery.

TABLE IVA

BLOOD OXYGEN SATURATIONS OF 7 PATIENTS IN WHOM ONLY SOME OF NECESSARY DATA WERE OBTAINED

No.	Reference	Percentage blood oxygen saturations						
		S.V.C.	R.A.	R.V.	Aorta	L.A.	L.V.	P.A.
31	M.S.	50	56	54	65	—	—	—
32	K.K.	40	42	58	65	—	—	—
33	R.B.	54	55	66	67	—	—	—
34	J.H.	54	55	70	70	—	—	—
35	Campbell (Case 1) ..	54	64	70	59	93	—	—
36	B.C.	54	66	70	64	91	—	—
37	K.M.	60	64	—	70	—	84	85

Abbreviations as in Table IA.

TABLE IVB

PRESSURES AND BLOOD FLOWS OF 7 PATIENTS IN WHOM ONLY SOME OF NECESSARY DATA WERE OBTAINED

No.	Blood pressures (mm. Hg)						Blood flows (l./min./m. ²)		
	Aorta	P.A.	R.V.	L.V.	R.A.	L.A.	Systemic	Pulmonary	Effective
31	110/85	—	—	—	2	—	4.8	—	1.4
32	69/49	—	69/3	—	2	—	4.5	—	1.9
33	120/75	—	120/10	—	3	—	4.0	—	1.2
34	100/65	—	100/0	—	2	—	5.0	—	1.7
35	—	—	—	—	—	—	6.32	74.5	0.93
36	89/50	P.S.	80/7	—	7	7	6.7	—	1.5
37	—	80/50	65/10	—	—	—	12.2	8.3	2.4

P.S.—Pulmonary stenosis; other abbreviations as in Table IA.

Group 2b (Table IIA and IIB). (Patients with a ventricular septal defect with the systemic blood flow exceeding the pulmonary blood flow, i.e. patients with severe pulmonary stenosis or high pulmonary vascular resistance.)

The right-to-left shunt: Left atrial and left ventricular blood samples were available in all the patients studied in this group. Seven patients (24, 25, 26, 27, 28, 29, and 30) showed evidence of a right-to-left shunt at atrial level; 3 of these (24, 25, and 26) showed evidence of a further right-to-left shunt at ventricular level, while in the remaining 4 (27, 28, 29, and 30) the saturation of the left atrial and left ventricular samples was similar. The right-to-left shunt in Patient 23 was at ventricular level.

The left-to-right shunt: No evidence of a left-to-right shunt at atrial level was found in this group. Right ventricular blood samples were available in only 6 patients: 3 of these (25, 28, and 29) showed an increase in the O₂ saturation in the right ventricle, while the blood samples from the right ventricle and the right atrium were similar in the remaining 3 patients (23, 24, and 27). The site of the left-to-right shunt in the last 3 patients could be directly from the left ventricle to the aorta through the ventricular septal defect, or from the pulmonary artery to the aorta through bronchial collaterals, provided pulmonary stenosis is not present.

The Inverse Relation of Pulmonary and Systemic Blood Flows. The inverse relation of the pulmonary and systemic blood flows in complete transposition of the great vessels occurs in cases associated with a large ventricular septal defect. Patients with a high pulmonary blood flow (low pulmonary vascular resistance or only mild pulmonary stenosis) tend to have a low systemic blood flow, and patients with a low pulmonary blood flow (severe pulmonary stenosis or high pulmonary vascular resistance) tend to have a high systemic blood flow. This is demonstrated by comparing

the pulmonary and systemic blood flows in Groups 2a and 2b, but is best shown in Patient 11, where the pulmonary blood flow was 18.0 l./min./m.² and the systemic blood flow was 4.4 l./min./m.², and Patient 27 where the pulmonary blood flow was 6.0 l./min./m.² and the systemic blood flow was 18.0 l./min./m.². The shunts in these two patients were 2.6 and 1.0 l./min./m.² respectively.

Pressures. *Group 1* (patients with a closed ventricular septum). The pressure in the left ventricle was lower than that in the right ventricle in 7 patients, the two pressures were equal in Patient 1 and the left ventricular pressure was higher than that of the right ventricle in Patient 7. The mean left atrial pressure was higher than the mean right atrial pressure in 5 patients, the two pressures were equal in 3 patients, and the mean right atrial pressure was higher than that of the left atrium in the remaining patient (No. 7). The 5 patients with a higher mean left atrial pressure had only a left-to-right shunt at atrial level. Patients 6, 8, and 9 with equal mean atrial pressures and Patient 7 with the mean right atrial pressure higher than that of the left atrium had a bidirectional shunt at atrial level.

Group 2 (patients with a ventricular septal defect). The systolic pressures in the two ventricles were within 5 mm. Hg of each other in all except Patients 18 and 27 in whom there was a difference of 15 and 10 mm. Hg respectively between the two ventricles. The pressure in the pulmonary artery, of course, depended on the presence or absence of pulmonary stenosis.

In Group 2a (13 patients) with a ventricular septal defect and the pulmonary blood flow exceeding the systemic flow, the mean left atrial pressure was higher than the mean right atrial pressure in 10 patients, the two pressures were equal in 1 (Patient 11), and the left atrial pressure was not available in the remaining 2 patients (21 and 22).

In Group 2b (8 patients) with a ventricular septal defect and the systemic blood flow exceeding the pulmonary blood flow, the mean right atrial pressure was higher than the mean left atrial pressure in 6, and the two pressures were equal in 2 (Patients 26 and 30).

DISCUSSION

Although the shunt in complete transposition of the great vessels, that is the effective pulmonary blood flow, is extremely limited, the average being 1.62 l./min./m.² in 37 patients, the actual flows in the pulmonary and systemic circuits varied from 1.7–18.0 l./min./m.² and 3.2–18.0 l./min./m.² respectively. This finding excludes the possibility that the shunt is responsible for the high flows in the pulmonary or systemic circuits, and neither does it support the suggestion of Astley and Parsons (1952), Wood (1956), Kjellberg *et al.* (1959), Taussig (1960), Edwards (1960), and others, that the volume and the direction of the shunt in transposition of the great vessels are probably determined by the total resistances in the two circuits. Had this been so, we would have expected that the shunt and consequently the pulmonary blood flow would increase in exactly the same proportions as the pulmonary vascular resistance drops, which is not the case. It appears, therefore, that the blood that goes around in the pulmonary or systemic circuits is made up largely of the output of the left or right ventricle respectively.

Several authors (Lenkei *et al.*, 1959; Keith *et al.*, 1958; Kjellberg *et al.*, 1959) have found that in cases of complete transposition of the great vessels, the two atrial pressures are often different. Noonan and her colleagues (1960) further noticed that in a group of patients with complete transposition of the great vessels associated with severe pulmonary stenosis and with the systemic blood flow exceeding the pulmonary blood flow, the mean right atrial pressure was often higher than the mean left atrial pressure. In another group of patients with low pulmonary vascular resistance, and the pulmonary blood flow exceeding the systemic blood flow, they noticed that the mean left atrial pressure was often higher than the mean right atrial pressure. This finding is interesting since it suggests that in complete transposition of the great vessels the atrial pressures, and probably the ventricular diastolic pressures also, are partly related to the amount of blood going through the atria and the ventricles. In other words, in complete transposition of the great vessels, an inverse relation probably exists between the mean atrial pressures and probably the ventricular diastolic pressures also, and their corresponding resistance. Therefore if the pulmonary blood flow exceeds

the systemic blood flow in a case of transposition of the great vessels with mild pulmonary stenosis or low pulmonary vascular resistances, and if an atrial septal defect and a ventricular septal defect are present, a left-to-right shunt may occur at atrial level, and a further left-to-right shunt at ventricular level may also occur during diastole. An equivalent right-to-left shunt will have to return to the pulmonary circuit and this probably will be during ventricular systole. The fact that, in 7 out of 9 patients with pulmonary blood flow exceeding the systemic blood-flow, the left ventricular sample was fully saturated, suggests that the right-to-left shunt, at least in some patients, could be directly from the right ventricle to the pulmonary artery through the ventricular septal defect, or from the aorta to the pulmonary artery through collateral vessels. In either of these conditions the right-to-left shunt must have left the right ventricle during systole only. If only a ventricular septal defect is present, a shunt from the left ventricle to the right ventricle probably occurs during diastole and an equal shunt from the right ventricle to the pulmonary circuit will probably occur during systole.

If the systemic blood flow exceeds the pulmonary flow, e.g. with severe pulmonary stenosis or with high pulmonary vascular resistance, the mean right atrial pressure, and probably the right ventricular diastolic pressure also, will exceed the corresponding pressures in the left side of the heart. A right-to-left shunt will occur probably at atrial level and a further right-to-left shunt at ventricular level may also occur during diastole. The left-to-right shunt has to be at ventricular or pulmonary artery level. In 3 out of 6 patients in this group, there was no evidence of a left-to-right shunt at ventricular level, indicating that this shunt must have occurred either directly from the left ventricle to the aorta through the ventricular septal defect or from the pulmonary artery to the aorta through anastomotic vessels if pulmonary hypertension was present. In either of these conditions the left-to-right shunt must have left the left ventricle during systole only. In the remaining 3 patients there was evidence of a left-to-right shunt at ventricular level.

The inverse relation of the pulmonary and systemic blood flows in complete transposition of the great vessels cannot be coincidental. A shunt cannot be responsible for its development, because if this were so, the shunt would have to be large, which is not the case, and it would also have to be predominant in one direction, which never occurs in complete transposition of the great vessels. The occurrence of high systemic blood flow in cases with severe pulmonary stenosis or with a very high pulmonary vascular resistance, is probably related to the development of tissue anoxia in these cases.

The mechanism of the exchange of blood in transposition of the great vessels is probably as follows: if the pulmonary vascular resistance is lower than the systemic, a hyperkinetic circulation develops in the pulmonary circuit. This results in a rise in the mean left atrial pressure above that in the right atrium, and probably also a rise in the left ventricular diastolic pressure above that in the right ventricle. If atrial and ventricular septal defects are present, a left-to-right shunt may occur at atrial level, and a further left-to-right shunt at ventricular level may occur during diastole. The sum of this left-to-right shunt probably returns to the pulmonary circuit during right ventricular systole. If only a ventricular septal defect is present, the shunt is probably from the left ventricle to the right ventricle during diastole and from the right ventricle to the pulmonary circuit during systole. If severe pulmonary stenosis is present or a very high pulmonary vascular resistance develops, the amount of blood flow in the pulmonary circuit is diminished and approaches that in the circuit. The gradient between the mean atrial pressures (or the diastolic pressures in the two ventricles if one had existed) will be diminished, thus reducing the size of the shunt. This will result in tissue anoxia. Peripheral systemic vasodilatation results with the development of a systemic hyperkinetic circulation. A rise in the mean right atrial pressure above that in the left atrium and probably also an increase in the right ventricular diastolic pressure above that in the left ventricle will occur. This results in a right-to-left shunt at atrial level and probably a further right-to-left shunt at ventricular level during diastole. A left-to-right shunt from the left ventricle to the systemic circuit probably occurs during systole. If a ventricular septal defect only is present, a right-to-left shunt probably occurs at ventricular level during diastole and an equal left-to-right shunt from the left ventricle to the systemic circuit probably occurs during systole. This mechanism of blood

shunting in transposition of the great vessels, apparently determined primarily by gradients of pressure difference created by the rate of blood flows in the two circuits and secondly by the size of the associated defects, explains the limited nature of the shunt in this condition. It must be pointed out that the occurrence of a large atrial septal defect will tend to equalize the pressures in the atria but the shunt will obviously continue. If also the diastolic filling resistances of the two ventricles are similar, a bidirectional blood shunt may occur at atrial level (Shaher and Johnson, 1963).

Kjellberg *et al.* (1959) suggested that respiration probably played a role in the mechanism of blood shunting in transposition of the great vessels and thought that respiration caused a greater variation in left atrial pressure than in right atrial pressure. They also added that there was a possibility of a change in the direction of the interatrial shunt during each cardiac cycle. Although the part played by these two factors cannot be assessed accurately, it seems unlikely that it is very important. The hypothesis that the shunt is dependent on these two factors offers no explanation for the inverse relation between the pulmonary and systemic flows in cases with a large ventricular septal defect. It cannot, however, be denied that the direction of the shunt may change during each cardiac cycle, since, rarely, there is evidence of a left-to-right shunt at atrial level despite a higher mean pressure in the right atrium than in the left atrium, e.g. Patient 7.

Angiocardiology in 6 patients with a ventricular septal defect with a selective dye injection into the right atrium in 2 (J. B. and D. T.) and into the right ventricle in 4 (G. W., J. H., M. A., and Y. S.) showed opacification of the pulmonary artery directly from the right ventricle in 4 during systole. The left ventricle did not opacify before the pulmonary artery in any of these 4 patients (Fig. 1 and 2), presumably owing to the high position of the ventricular septal defect. This demonstrated that the right-to-left shunt in these 4 patients occurred during systole so that the opaque medium passed directly from the right ventricle to the pulmonary artery. In the fifth patient both the left ventricle and pulmonary artery opacified simultaneously from the right ventricle during systole, suggesting that the ventricular septal defect was either very large, as it appeared on the angiocardiology, or low in position. The sixth patient (Y.S.), had the clinical features of a hyperkinetic systemic circulation. The dye was seen to enter the left ventricle from the right ventricle during diastole (Fig. 3), suggesting that in this patient the right-to-left shunt occurred during diastole. Although the position of the tip of the catheter was away from the ventricular septal defect, reflux of the dye into the left ventricle during the injection cannot be excluded.

According to Abbott (1936), Astley and Parsons (1952), and Taussig (1960), the mechanism of blood shunting in complete transposition of the great vessels with a closed ventricular septum but with an atrial septal defect and patent ductus arteriosus depends on the presence of pulmonary hypertension, which makes possible a shunt from the pulmonary artery to the aorta through a patent ductus arteriosus; a balancing shunt from the right atrium to the left atrium is then postulated. If this were so, differential cyanosis would result, the upper part of the body being more blue than the lower part. Among the 50 patients reported by Noonan and her colleagues (1960), only one had differential cyanosis; the upper half of the body being more cyanosed than the lower half. On the other hand, Fowler and Orway (1952) and Keith and his colleagues (1958) found that patients with a closed ventricular septum usually had a left-to-right shunt at atrial level, while Fowler and Ordway did not see the clinical picture of differential cyanosis amongst their patients with transposition. If the upper half of the body including the brain and the coronary arteries were to be perfused with blood having low oxygen saturation, survival for a long time is not possible. To permit survival, a left-to-right shunt occurs at atrial level and for this to happen the pulmonary vascular resistance is usually lowered. These cases that lack a large ventricular septal defect or a large patent ductus arteriosus do not usually maintain the fetal high pulmonary vascular resistance; this results in a hyperkinetic pulmonary circulation which in turn leads to the development of a pressure gradient between the left and the right atria. Furthermore, if the left ventricular and pulmonary arterial pressures drop below the right ventricular and aortic pressures another gradient is created between the aorta and the pulmonary artery, permitting an effective right-to-left shunt, i.e. from the aorta to the pulmonary artery, through a patent ductus arteriosus or anastomotic vessels.

FIG. 1.—Lateral angiogram of patient J.B. Selective dye injection into the right atrium showing opacification of the right atrium (RA), right ventricle (RV), aorta (Ao), and pulmonary artery (PA). The left ventricle did not opacify.

FIG. 2.—Lateral angiogram of patient G. W. Dye injection into the right ventricle (RV) showing the anteriorly situated aorta (Ao), and the entry of the dye through a ventricular septal defect (VSD) to the posteriorly situated pulmonary artery (PA).

Two babies (M.P. and R.L.), in whom the diagnosis of transposition of the great vessels with a closed ventricular septum, a patent ductus arteriosus, and an atrial septal defect was confirmed at necropsy, were seen at Guy's Hospital. Both of them had been previously investigated by selective right atrial angiography (Fig. 4, 5, and 6). This demonstrated, in both, immediate filling of the right ventricle and the ascending aorta. Neither the left atrium nor the left ventricle of either

FIG. 3.—Lateral angiogram of patient Y.S. who had the clinical features of a hyperkinetic systemic circulation. The dye that was injected into the right ventricle (RV) has opacified the anteriorly situated aorta (Ao) and, through the ventricular septal defect, the left ventricle (LV) during diastole.

FIG. 4.—Lateral angiogram of patient M.P. who was shown at necropsy to have a closed ventricular septum, an atrial septal defect, and a patent ductus arteriosus. The dye was injected into the right atrium (RA) from which the right ventricle (RV) and aorta (Ao) opacified. Neither the left atrium nor the left ventricle opacified. The pulmonary artery (PA) opacified from the aorta through a patent ductus arteriosus, although the latter was not certainly shown in the angiogram.

patient opacified, but their pulmonary arteries were seen to opacify presumably through the ductus arteriosus although the latter was not definitely shown on the angiogram.

Patients with a closed ventricular septum and only a large atrial septal defect, have a bidirectional shunt at atrial level. The haemodynamics of these cases are probably similar to those of common atrium with direct mixing of the two blood-streams at atrial level.

The occurrence of increased systemic blood flow in complete transposition of the great vessels has been noticed by several authors (Campbell and Suzman, 1951; Cleland *et al.*, 1957; Noonan *et al.*, 1960; D. C. Deuchar, 1962, personal communication). In a group of 8 severely cyanosed patients with complete transposition of the great vessels associated with pulmonary stenosis (one of whom had

FIG. 5.—Postero-anterior angiogram of patient R.L. who was shown at necropsy to have a closed ventricular septum, an atrial septal defect, and a patent ductus arteriosus. The dye was injected into the right atrium (RA) from which the right ventricle (RV) and the aorta (Ao) opacified. Neither the left atrium nor the left ventricle opacified.

FIG. 6.—Lateral angiogram of patient R.L. with dye injection into the right atrium (RA) showing opacification of the right ventricle (RV) and aorta (Ao). The pulmonary artery (PA) opacified from the aorta presumably through a patent ductus arteriosus, although the latter was not certainly shown in the angiogram.

single ventricle), Cleland and his colleagues (1957) noticed that the peripheral pulses had a remarkably full volume and in one case there was visible pulsation of the dorsalis pedis and posterior tibial vessels. Noonan and her colleagues (1960) noticed that infants with marked cyanosis had varicosities of the extremities and scalp. Patients with a hyperkinetic systemic circulation either have small associated defects, e.g. a small ventricular septal defect or a closed ventricular septum, or a large ventricular septal defect with severe pulmonary stenosis or a high pulmonary vascular resistance. Clinically these patients are usually extremely cyanosed and may have pounding pulses, distended finger and arm veins, and warm extremities. Their electrocardiograms may show severe right ventricular hypertrophy and strain, but tall P waves may be present especially in lead II, as in Patient Y.S.

SUMMARY

The hæmodynamics of 37 patients with complete transposition of the great vessels were studied: 26 patients were collected from the published reports, and the remaining 11 patients were seen at Guy's Hospital and at Southampton Chest Hospital. Although the shunt was small in each and averaged 1.62 l./min./m.², the actual pulmonary and systemic blood flows varied within a much wider range. An inverse relation exists between the pulmonary and the systemic blood flows in cases with a large ventricular septal defect. Patients with a low pulmonary blood flow, or who have small associated defects, develop a hyperkinetic systemic circulation. It is suggested that the mechanism of the exchange of blood in complete transposition of the great vessels is initiated by gradients between the mean atrial pressures and probably also the diastolic ventricular pressures, created by the rate of blood flow in the two circuits. The amount of blood gained by one circuit in this way is likely to be returned to the original circuit during ventricular systole.

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