Acid-base Balance in Complete Transposition of the Great Vessels

REDA M. SHAHER* AND LANGFORD KIDD*

From the Department of Paediatrics, University of Toronto, and the Research Institute of the Hospital for Sick Children, Toronto, Ontario, Canada

In complete transposition of the great vessels the pulmonary and systemic circulations exist as two separate circuits. Although the pulmonary and the systemic blood flows may vary widely, the effective pulmonary blood flow is often small (Shaher, 1964). While the acid-base balance in the systemic circuit has been studied by several workers (Sunico, Harned, and Ordway, 1960; Gootman, Scarpelli, and Rudolph, 1963; Talner *et al.*, 1965), there is very little known about the acid-base balance in the pulmonary circuit (Gootman *et al.*, 1963) and the relation between them. The object of this paper is to present the findings in a group of patients with complete transposition of the great vessels in all of whom the acid-base balance has been determined in both the pulmonary and the systemic circuits. The large majority of these patients had associated defects, either congenital or surgically created, and had survived beyond infancy.

SUBJECTS AND METHODS

Nineteen patients with complete transposition of the great vessels were investigated by cardiac catheterization. They were divided into three groups as follows:

Group I: eight patients with an intact ventricular septum.

Group II: seven patients with a ventricular septal defect.

Group III: four patients with a ventricular septal defect and pulmonary stenosis.

Their ages at cardiac catheterization ranged from 1 day to 13 years. An atrial septal defect had been surgically created earlier in life in 10, a Blalock-Taussig anastomosis

performed in one, and a Baffe's operation in one. Mild sedation (Smith, Rowe, and Vlad, 1958) before cardiac catheterization was given to all patients except those in the first two months of life. All patients were quiet and breathing room air. Blood samples were taken from the aorta (Ao) and a pulmonary vein (PV) in each case and, in addition, from the superior vena cava (SVC) and left ventricle (LV) or pulmonary artery (PA) in 10. Samples were taken anaerobically in rapid succession into heparinized syringes and were analysed within five minutes for pH, Po₂ and PCO₂. pH was measured using a micro-glass electrode, Po₂ using a Clark-type electrode, and PCO₂ using a modified Severinghaus electrode (Instrumentation Laboratories, Inc.). The electrodes were calibrated between each estimation using known gas mixtures. In addition, blood lactate and pyruvate levels were measured in systemic arterial blood in 10 of these 19 patients, using the enzymatic methods described by Scholz *et al.* (1959), and by Hess (1955) (Calbiochem). The normal ranges for this method in our laboratory are for pyruvate 0.2 to 0.7 mg./100ml. (0.02 to 0.08 mEq/l.) and for lactate 3–11 mg./100 ml. (0.33 to 1.22 mEq/l.). In calculating pulmonary, systemic, and effective pulmonary blood flows the following equations have been used:

$$\text{Pulmonary blood flow} = \frac{\text{O}_2 \text{ consumption}}{\text{PV O}_2 \text{ content} - \text{PA O}_2 \text{ content}}$$

$$\text{Systemic blood flow} = \frac{\text{O}_2 \text{ consumption}}{\text{Ao O}_2 \text{ content} - \text{SVC O}_2 \text{ content}}$$

$$\text{Effective blood flow} = \frac{\text{O}_2 \text{ consumption}}{\text{PV O}_2 \text{ content} - \text{SVC O}_2 \text{ content}}$$

The oxygen consumption has been assumed to be 180 ml./min.m². In calculating the pulmonary blood flow in cases where the pulmonary arterial blood sample has not been obtained, the left ventricular blood sample has been used instead. In all cases the diagnosis was confirmed by angiocardiography.

Received December 17, 1965.

* Aided by a grant from the Ontario Heart Foundation and the Department of National Health and Welfare, Ottawa, Canada.

TABLE I
BLOOD pH, Po₂, AND Pco₂ OF 8 PATIENTS WITH INTACT VENTRICULAR SEPTUM

Patient No. and age	pH				Po ₂ (mm. Hg)				Pco ₂ (mm. Hg)				Comments
	SVC	Aorta	Pulmonary vein	LV-PA	SVC	Aorta	Pulmonary vein	LV-PA	SVC	Aorta	Pulmonary vein	LV-PA	
1 1 dy.	—	7.26	7.37	—	—	20	—	—	—	35	21	—	ASD created at 1 month ASD created at 2 weeks ASD created at 1 week Blalock-Taussig, at 4 years Baffle's operation at 6 years
2 5 dy.	—	7.36	7.44	—	—	18	108	—	—	33	26	—	
3 1 yr. 11 mth.	—	7.37	7.39	—	—	51	100	—	—	38	35	—	
4 1 yr. 2 mth.	7.35	7.38	7.39	7.37	28	40	96	53	40	36	31	34	
5 1 yr. 4 mth.	7.37	7.41	7.44	—	30	38	114	—	46	43	30	—	
6 1 yr. 9 mth.	7.40	7.37	7.41	7.38	32	41	94	54	46	42	37	39	
7 8 yr. 10 mth.	—	7.36	7.39	—	—	27	109	—	—	31	29	—	
8 13 yr. 1 mth.	7.368	7.385	7.395	7.375	37	44	93	51	37	34	32	35	
Mean	7.37	7.35	7.40	7.375	32	35	120	53	42	36.5	30	36	

SVC=superior vena cava; LV=left ventricle; PA=pulmonary artery.

TABLE II
BLOOD pH, Po₂, AND Pco₂ OF 7 PATIENTS WITH A VENTRICULAR SEPTAL DEFECT

Patient No. and age	pH				Po ₂ (mm. Hg)				Pco ₂ (mm. Hg)				Comments
	SVC	Aorta	Pulmonary vein	LV-PA	SVC	Aorta	Pulmonary vein	LV-PA	SVC	Aorta	Pulmonary vein	LV-PA	
9 6 mth.	—	7.32	7.42	7.40	—	37	99	58	—	43	29	37	ASD created at 3 weeks
10 11 mth.	—	7.40	7.42	7.43	—	41	100	57	—	33	26	30	ASD created at 2 months
11 1 yr. 2 mth.	7.30	7.29	7.35	7.30	35	40	79	54	47	45	37	42	ASD created at 3 months
12 1 yr. 3 mth.	7.42	7.45	7.48	7.44	33	42	96	64	41	38	31	30	
13 2 yr.	—	7.38	7.41	—	—	38	91	—	—	47	34	—	ASD created at 4 months
14 2 yr. 11 mth.	7.31	7.30	7.44	—	31	34	82	—	36	38	28	—	
15 5 yr. 10 mth.	7.20	7.39	7.40	7.38	35	36	93	61	36	37	31	34	ASD created at 4½ years
Mean	7.307	7.36	7.41	7.39	33.5	38	91	59	40	40	31	34	

Abbreviations as in Table I.

TABLE III
BLOOD pH, Po₂, Pco₂ OF 4 PATIENTS WITH A VENTRICULAR SEPTAL DEFECT AND PULMONARY STENOSIS

Patient No. and age	pH				Po ₂ (mm. Hg)				Pco ₂ (mm. Hg)				Comments
	SVC	Aorta	Pulmonary vein	LV-PA	SVC	Aorta	Pulmonary vein	LV-PA	SCV	Aorta	Pulmonary vein	LV-PA	
16 8 mth.	—	7.365	7.415	7.395	—	14	70	43	—	28.5	17	24.5	ASD created at 6 months
17 1 yr. 2 mth.	—	7.40	7.50	—	—	40	102	—	—	37	26	—	
18 3 yr. 9 mth.	—	7.38	7.46	—	—	24	93	—	—	45	30	—	
19 3 yr. 9 mth.	—	7.35	7.38	7.37	—	41	84	56	—	40	34.3	35.5	
Mean	—	7.37	7.44	7.38	—	30	87	49.5	—	37.5	27	30	

Abbreviations as in Table I.

RESULTS

Acid-base Balance. The results are given in Tables I, II, and III. It will be seen that in all three groups the pH of the systemic blood varied on the acid side of normal, while that from the pulmonary circuit was more alkalotic. In all three groups the Pco₂ of the systemic blood was considerably higher than that in the pulmonary circulation. The mean pH, Pco₂, and Pco₂, as well as the standards of deviation in the SVC, Ao, PV, and PA or LV, are illustrated in Fig. 1, 2, and 3.

Lactates and Pyruvates. The results are given in Table IV. It can be seen that the results were within normal limits except in Patients 1 and 2. These were the youngest and were clinically the most severely ill.

Haemodynamics (Table V). In Group I the systemic arterial O₂ saturation varied from 21 to 76 per cent, with an average of 62 per cent. The pulmonary blood flow exceeded the systemic blood flow in the five patients in whom this measurement

FIG. 1.—Mean pH in the pulmonary and the systemic circuits in 19 patients with transposition of the great vessels. Vertical lines represent standard derivations: SVC, 7.34 ± 0.022 ; Ao, 7.36 ± 0.014 ; PV, 7.42 ± 0.025 ; PA-LV, 7.39 ± 0.012 (abbreviations as in Table I).

FIG. 3.—Mean PCO₂ in the pulmonary and the systemic circuits in 19 patients with transposition of the great vessels. Vertical lines represent standard derivations: SVC, 41.1 ± 4.3 ; Ao, 38.4 ± 4.7 ; PV, 29.7 ± 5.0 ; PA-LV, 34.2 ± 4.7 .

was available. In Group II the systemic arterial saturation varied from 53 to 73 per cent, with an average of 65 per cent. Apart from Patient 11, in whom the pulmonary blood flow equalled the systemic blood flow, in the remaining six patients the pulmonary blood flow exceeded the systemic blood flow. In Group III the systemic arterial saturation varied from 38 to 70 per cent, with an average of 53 per cent. In Patient 17, the pulmonary blood flow slightly exceeded the systemic flow, while in the remaining three patients the

FIG. 2.—Mean PO₂ in the pulmonary and the systemic circuits in 19 patients with transposition of the great vessels. Vertical lines represent standard derivations: SVC, 32.6 ± 2.8 ; Ao, 35.1 ± 9.6 ; PV, 94.6 ± 10.7 ; PA-LV, 55.1 ± 5.4 .

TABLE IV

BLOOD LACTATES AND PYRUVATES IN 10 PATIENTS WITH TRANSPOSITION OF THE GREAT VESSELS

Patient No.	Lactate (mg./100 ml.)	Pyruvate (mg./100 ml.)	Ratio	Po ₂
1	38.25	1.858	20.61	20
2	92.022	3.371	27.3	18
3	6.00	0.410	14.6	51
5	5.051	0.537	9.4	38
6	6.063	0.476	12.9	41
8	6.563	0.330	19.9	44
9	8.313	0.641	12.7	37
11	3.250	0.378	8.4	40
15	6.313	0.376	16.7	36
19	8.813	0.732	12.0	41

Note: Samples are from the systemic artery.

pulmonary blood was either equal to or less than the systemic blood flow. Apart from Patient 18, the mean left atrial pressure in all remaining 18 patients was normal.

TABLE V
ESSENTIAL HAEMODYNAMIC DATA

Patient No.	Mean left atrial pressure	Systemic arterial saturation	Effective pulmonary blood flow (litres/min./m ²)	Pulmonary blood flow (litres/min./m ²)	Systemic blood flow (litres/min./m ²)
1	8	21	0.9	—	7.0
2	5	33	1.1	—	5.0
3	5	79	2.1	12.2	3.7
4	4	70	1.68	5.8	3.9
5	2	67	1.8	9.0	4.7
6	8	70	2.4	9.0	5.0
7	2	57	1.1	7.0	5.0
8	4	76	2.0	5.8	3.9
9	8.5	53	1.4	8.2	4.6
10	9	73	2.1	20.0	3.6
11	5	64	2.0	7.0	7.0
12	7.5	65	1.5	11.0	3.2
13	5	66	1.4	7.5	3.2
14	5	62	1.3	7.5	4.0
15	2	70	1.3	4.5	2.6
16	4	40	1.4	6.0	14.0
17	6	70	1.7	5.5	3.7
18	13	38	1.9	8.5	12.0
19	7	63	1.3	4.4	4.7

DISCUSSION

While the occurrence of systemic acidosis in cyanotic congenital heart disease is well recognized, the information available concerning transposition of the great vessels is limited. Sunico *et al.* (1960) measured the acid-base balance in the systemic circuit of 14 patients with complete transposition of the great vessels, securing, however, a sample from the pulmonary vein in only one of them. Gootman *et al.* (1963) more recently reported a study in four patients with cyanotic congenital heart disease, one of whom had transposition of the great vessels: all had in common a conspicuously reduced effective pulmonary blood flow, and pulmonary venous blood samples were obtained in each case.

The present work increases the data available by the addition of measurements of acid-base balance in both systemic and pulmonary circulations in a further 19 patients with complete transposition of the great vessels. It is clear that in each of our patients, regardless of the anatomy of the associated defects or the amount of pulmonary blood flow, the systemic arterial sample had a lower pH and a higher PCO₂ than that obtained from the pulmonary veins. From the point of view of the pulmonary venous blood, therefore, hyperventilation is implied. Since the systemic arterial PCO₂ and pH in complete transposition result from the admixture of a large amount of systemic venous blood with a low pH and PO₂ and high PCO₂ with a small amount of pulmonary venous blood (effective flow) with a high pH and PO₂ and low PCO₂, which has crossed through the associated defects, the hyperventilation helps to maintain the systemic arterial acid-base status

normally or only slightly towards the acid side. The hyperventilation may thus be considered as a barely effective respiratory compensation for systemic acidosis. It is similarly obvious that the pulmonary arterial PCO₂ and pH result from the admixture of a large amount of pulmonary venous blood with a small amount of systemic venous blood which has crossed through the associated defects.

Huckabee (1958) has shown that anaerobic tissue metabolism begins to assume significance at systemic arterial PO₂ levels below 35 mm. Hg. At these levels the lactic acid-pyruvic acid relationship is disturbed, resulting in a greater proportion of lactic acid. This is supported by the present series in that Patients 1 and 2 with the lowest arterial PO₂ had the highest lactates and lactate/pyruvate ratios.

Recently Talner and his co-workers (1965) studied the acid-base balance in the systemic circuit of 20 patients with congenital heart disease who were in heart failure. Three of them had transposition of the great vessels. They concluded that the occurrence of systemic acidosis in these patients must be respiratory in origin, since all the patients they studied shared the features of large pulmonary blood flow and pulmonary congestion. They suggested that pulmonary congestion, giving rise to diminished lung compliance and airway obstruction, produced respiratory acidosis.

The data from the present series of 19 patients show that the pulmonary venous PCO₂ was low in all of them, suggesting that there was no abnormality in the gaseous exchange mechanisms at alveolar-capillary level. In addition, pulmonary venous congestion, as evidenced by mean left atrial pressure, was absent in 18 of our 19 patients.

Patient 18, with mildly raised left atrial pressure, had pulmonary stenosis. Pulmonary venous congestion and heart failure, however, may be responsible for the abnormalities in the other patients described by Talner, and certainly heart failure might be expected to increase the abnormalities in the systemic circuit in patients with complete transposition of the great vessels through the mechanism described.

Lastly, it is clear that our patients are not truly representative of the entire range of cases with transposition of the great vessels, a large number of whom present early in life with a much more severe disturbance than is demonstrated here. However, for obvious reasons it is not always possible to secure full information regarding acid-base status in both circulations in these severely ill infants. These data present the picture in those children who, because of associated defects—either surgical or congenital—and presumably larger effective pulmonary blood flow, have survived infancy. The disturbance in the severely ill group is likely to be of the same nature but more grave (i.e. with more acidosis in the systemic and more alkalosis in the pulmonary circulation), because of lack of sufficient communication between the two circulations, and complicated, as in two of our patients, by a lactic acidosis due to hypoxia.

SUMMARY

Blood samples were taken from pulmonary vein and systemic artery in 19 children with complete

transposition of the great vessels, and were analysed for oxygen and carbon dioxide tensions and pH. Of the 19, 11 had a ventricular septal defect (with pulmonary stenosis in four), while eight had an intact ventricular septum. In all of them the pulmonary vein PCO₂ was lower and the pH higher than in the aorta. Lactate/pyruvate ratio was measured in 10 out of 19 patients and was highest in the two who had the lowest systemic arterial saturation.

REFERENCES

- Gootman, N. L., Scarpelli, E. M., and Rudolph, A. M. (1963). Metabolic acidosis in children with severe cyanotic congenital heart disease. *Pediatrics*, **31**, 251.
- Hess, B. (1955). Bemerkungen zur Bestimmung der Brenztraubensäure im menschlichen Serum. *Klin. Wschr.*, **33**, 540.
- Huckabec, W. E. (1958). Relationship of pyruvate and lactate during anaerobic metabolism. III. Effect of breathing low-oxygen gases. *J. clin. Invest.*, **37**, 264.
- Scholz, R., Schmitz, H., Bücher, T., and Lampen, J. O. (1959). Über die Wirkung von Nystatin auf Bäckerhefe. *Biochem. Z.*, **331**, 71.
- Shaher, R. M. (1964). The haemodynamics of complete transposition of the great vessels. *Brit. Heart J.*, **26**, 343.
- Smith, C., Rowe, R. D., and Vlad, P. (1958). Sedation of children for cardiac catheterization with an ataractic mixture. *Canad. Anaesth. Soc. J.*, **5**, 35.
- Sunico, R. M., Harned, H., Jr., and Ordway, N. K. (1960). Respiratory acidosis in transposition of the great vessels. *Amer. J. Dis. Child.*, **100**, 531.
- Talner, N. S., Sanyal, S. K., Halloran, K. H., Gardner, T. H., and Ordway, N. K. (1965). Congestive heart failure in infancy. I. Abnormalities in blood gases and acid base-equilibrium. *Pediatrics*, **35**, 20.