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*From the Medical Clinic of Prof. Mikamo, School of Medicine, University of Tokyo
(Director: Professor Y. Mikamo, M. D.)*

Studies on the Circulatory Dynamics in Congenital Heart Disease

By

MASAMITSU SHIBA

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PREFACE

Recently the use of cardiac catheterization technique in clinical cardiology has greatly contributed especially for the authentic diagnosis of congenital heart disease, of which we are now able to study even detailed circulatory dynamics¹⁻⁵. In the Medical Clinic of Prof. Mikamo at Tokyo University, more than one hundred patients with cardiovascular anomalies were catheterized in the last several years, of whom the seventy-seven cases were reviewed in this paper under a classification into several anatomic-physiological categories, especially with a view to revealing (1) some hemodynamic principles generally operating in the lesser circulatory system under certain types of congenital heart disease, and (2) several physiologic significances concerned with pulmonary stenosis particularly in the tetralogy of Fallot and some related anomalies.

MATERIAL AND METHODS

Case histories of seventy-seven patients of congenital heart disease were collected, whose diagnosis were all ascertained through cardiac catheterizations, and almost all of them were regarded as to be free from severe cardiac insufficiency. The catheterization was carried out in the usual fashion for the patients resting in a fairly basal metabolic state. Blood samples were obtained through the catheter and by a direct puncture of the femoral or the brachial artery, during which the expired air was collected in a Douglas bag. Samples were analyzed for oxygen content, capacity and saturation by the methods of Van Slyke and Neill⁶. Pressures were recorded with Hamilton manometers⁷ or in the

later part of the series with capacitance manometer⁸), and mean pressures were checked with a saline manometer. Brachial arterial pressures were measured sphygmomanometrically and calculated for mean pressures by the formula of Wezler and Böger⁹).

Systemic, pulmonary, and "effective pulmonary" blood flows were calculated according to the methods described by Bing and associates²), based originally upon Fick's principle. Note here that the "effective pulmonary" flow means the pulmonary flow of mixed venous blood, so the difference of which to the systemic or pulmonary flow corresponds to the overall amount of the right-to-left or left-to-right shunt respectively. In cases with multiple shunts in a same heart the Armstrong's method was employed to estimate the individual ones¹⁰). Vascular resistances in the systemic and pulmonary systems were calculated using the methods described by Bayer et al.⁵), derived principally from Poiseill's law. As for estimations of ventricular works the author obeyed to the formulas offered by Gorlin and associates¹¹), but using the right ventricular mean pressure in place of the pulmonary arterial mean pressure for the work of right ventricle¹²). Stenotic pulmonary valvular mean area (cm²) was estimated at a quotient of the mean pulmonary flow (cc/sec) divided by the square root of the mean pressure difference (mmHg) of the right ventricle and the pulmonary artery, as suggested by Rodrigo and Snellen¹³), among others¹⁴). Thus calculated areas, however, are not always identical with the actual pulmonary orifice areas apart from those in case of the pure valvular stenosis, because in this method the so-called contraction factor, that is the rate of energy loss across

the stenotic opening, is prescribed to 1.0 notwithstanding variable anatomic types of the stenosis. Accordingly, in the strict sense, they may be rather designated as "mean effective pulmonary valve

areas", under the character of a pertinent index of the "physiological" pulmonary stenosis. Similarly, the "stenotic pulmonary valve resistance" in this paper, calculated according to the method

TABLE I (a) CATHETERIZATION RESULTS OF THE SEVENTY-SEVEN PATIENTS OF CONGENITAL HEART DISEASE.

Case No.	Name	Age Sex	BSA	O ₂ Cons.	Art. O ₂ Sat.	O ₂ Cap.	A.-V. O ₂ Difference				Blood Flow			Shunt		Mean Pressure					* Vascular Resistance			Ventricular Work			** PVMA (DA)			
							Sys Pul Eff	Sys Pul Eff	Sys Pul Eff	Sys Pul Eff	L-R	R-L	RA	LA	RV	LV	PA	BA	Sys Pul SPVR	R V LV	R/L									
yr. m ² cc/min/m ² % vol. % lit./min./m ² mm. Hg dynes sec.cm ⁻⁵ kgM/min/m ² % cm ²																														
P.D.A. - Type I.																														
1	Y.A.	3½F	1.3	155	94	17.5	3.6	3.1	3.6	4.3	5.1	4.3	0.8	0	0	-	8	-	12	84	997	110	-	0.8	4.9	17	-	-	-	
2	M.O.	25F	1.4	157	95	16.8	3.6	3.4	3.6	4.3	4.6	4.3	0.3	0	3	-	13	-	15	85	905	170	-	0.6	5.2	10	-	-	-	
3	K.H.	20F	1.4	165	92	19.4	4.0	3.0	4.0	4.1	5.5	4.1	0.9	0	3	-	10	-	15	81	841	146	-	0.7	6.4	17	(0.2)	-	-	
4	S.U.	22F	1.5	137	92	16.0	2.9	2.7	2.9	4.7	5.1	4.7	0.4	0	3	-	12	-	19	65	1120	219	-	0.9	4.4	22	(0.1)	-	-	
5	A.I.	15M	1.6	139	95	19.5	3.1	1.6	3.1	4.5	8.7	4.5	2.1	0	4	-	13	-	17	74	976	124	-	0.8	6.2	26	(0.4)	-	-	
Mean	23	1.4	150	94	17.5	3.4	2.8	3.4	4.4	5.8	4.4	0.9	0	3	-	11	-	16	78	984	154	-	0.8	5.2	16	(0.2)	-	-	-	
s.d.	-	-	11	1	1.4	0.4	0.6	0.4	0.2	1.7	0.2	0.7	0	1	-	2	-	2	7	116	38	-	0.3	0.6	6	-	-	-	-	
P.D.A. - Type II.																														
6	M.O.	18F	1.2	168	96	16.0	3.5	2.3	3.5	4.9	7.3	4.9	2.5	0	7	-	12	-	20	81	781	164	-	1.2	7.9	24	(0.5)	-	-	
7	M.S.	6M	0.7	184	95	17.2	4.2	2.5	4.2	4.4	7.5	4.4	3.0	0	7	-	17	-	22	70	900	182	-	1.5	7.0	33	-	-	-	
8	Y.K.	13F	1.3	159	97	16.0	3.9	1.6	3.9	4.2	10.1	4.2	5.9	0	6	-	15	-	22	53	1748	135	-	1.0	6.0	17	-	-	-	
9	Y.T.	33F	1.2	154	92	18.1	3.4	2.2	3.4	4.7	7.1	4.7	2.4	0	6	-	22	-	26	103	1160	236	-	1.3	10.3	13	-	-	-	
10	A.N.	26F	1.0	171	94	17.0	3.7	1.7	3.7	4.7	10.4	4.7	5.7	0	3	-	16	-	26	73	1586	162	-	1.5	8.7	18	(0.8)	-	-	
11	M.T.	9M	0.9	154	86	16.7	3.9	2.1	3.9	3.9	7.4	3.9	3.5	0	4	-	20	-	27	77	1555	238	-	1.3	7.7	18	(0.4)	-	-	
Mean	18	1.1	165	94	16.8	3.8	2.1	3.8	4.5	8.3	4.5	3.8	0	5	-	17	-	24	76	1288	186	-	1.3	7.5	18	(0.6)	-	-	-	
s.d.	-	-	11	3	0.7	0.3	0.5	0.3	0.3	1.4	0.3	1.4	0	1	-	3	-	3	15	316	39	-	0.5	1.2	7	-	-	-	-	
P.D.A. - Type III.																														
12	T.S.	7M	0.7	166	99	12.1	4.4	1.9	4.4	4.4	7.5	4.4	3.0	0	4	-	21	-	35	60	1040	322	-	1.6	6.7	24	(0.9)	-	-	
13	H.F.	11F	1.1	161	94	21.2	6.3	2.5	6.3	2.5	6.4	2.5	3.9	0	3	-	28	-	42	64	2185	460	-	1.4	3.4	46	(1.0)	-	-	
14	T.H.	18M	1.4	159	92	20.4	5.0	2.5	5.0	3.2	6.3	3.2	3.1	0	7	-	30	-	56	119	728	646	-	2.2	10.3	22	(0.5)	-	-	
15	M.N.	4F	0.5	184	74	15.6	6.2	5.0	6.2	3.0	3.7	3.0	1.3	0.3	9	-	44	-	50	55	1143	965	-	1.8	3.5	50	(0.5)	-	-	
Mean	10	0.9	168	90	17.3	5.8	3.0	5.8	3.3	6.0	3.3	2.9	-	6	-	31	-	46	75	1274	598	-	1.8	6.0	36	(0.9)	-	-	-	
s.d.	-	-	10	9	3.7	0.8	1.2	0.8	0.7	2.8	0.7	1.0	-	2	-	8	-	8	26	549	241	-	0.3	2.8	12	-	-	-	-	
A.S.D. - Type I.																														
16	I.T.	29F	1.2	166	98	16.9	4.2	3.4	4.2	3.9	4.8	3.9	0.9	0	2	-	12	-	10	82	1588	83	-	0.8	4.4	19	-	-	-	
17	R.S.	29M	1.4	110	94	15.4	3.0	2.0	3.0	3.7	5.4	3.7	1.7	0	2	-	12	-	12	75	1566	103	-	1.1	5.3	20	-	-	-	
18	K.M.	25F	1.8	178	94	16.8	4.3	2.6	4.3	4.1	6.6	4.1	2.5	0	5	-	12	-	16	84	1501	132	-	1.2	4.7	25	-	-	-	
19	S.T.	26M	1.5	162	93	16.1	3.7	2.2	3.7	3.4	5.0	3.4	1.6	0	7	-	14	-	16	77	1634	177	-	1.1	3.5	30	-	-	-	
20	M.K.	20M	1.7	148	95	19.0	3.5	2.7	3.5	4.0	5.4	4.0	1.4	0	3	-	15	-	20	91	1726	226	-	1.2	4.9	24	-	-	-	
21	K.O.	26F	1.5	151	95	17.0	4.5	1.9	4.5	3.4	8.2	3.4	4.8	0	3	-	16	-	20	93	2104	147	-	1.9	5.2	36	-	-	-	
22	M.T.	31M	1.5	182	94	20.8	5.8	2.3	5.8	3.2	7.9	3.2	4.7	0	4	-	21	-	23	82	1957	181	-	1.6	3.5	45	-	-	-	
23	R.N.	20F	1.5	157	96	20.6	3.6	1.4	3.6	4.4	11.5	4.4	7.0	0	6	-	24	-	27	87	1456	154	-	2.3	5.2	44	-	-	-	
24	R.T.	16M	1.3	153	95	17.0	5.1	1.3	5.1	3.0	12.1	3.0	9.0	0	12	-	24	-	40	89	2040	233	-	4.5	3.8	120	-	-	-	
Mean	25	1.5	156	95	17.7	4.0	2.2	4.0	3.7	7.4	3.7	3.7	0	5	-	10	-	18	-	20	85	1753	159	-	1.7	4.5	40	-	-	-
s.d.	-	-	20	1	1.8	0.8	0.5	0.8	0.5	3.5	0.5	2.6	0	3	-	8	-	8	7	231	47	-	1.1	0.7	30	-	-	-	-	
A.S.D. - Type II.																														
25	K.A.	18F	1.3	160	92	15.8	3.9	3.4	4.4	4.1	5.3	3.6	1.7	0.5	4	-	15	-	18	77	1428	192	-	1.3	4.3	31	-	-	-	
26	M.U.	32M	1.5	170	89	21.7	4.1	2.2	5.4	4.1	7.9	3.2	4.8	1.0	7	-	15	-	17	77	1330	121	-	1.7	4.2	41	-	-	-	
27	T.A.	20M	1.7	178	90	20.9	4.5	2.8	5.6	4.0	6.4	3.2	3.3	0.8	7	-	15	-	21	88	1608	199	-	1.4	4.8	29	-	-	-	
28	M.O.	19M	1.6	146	90	21.9	3.2	2.0	4.0	4.6	7.4	3.7	3.7	1.0	6	-	15	-	20	83	1330	190	-	1.6	5.2	37	-	-	-	
29	N.O.	22F	1.4	152	90	15.5	4.3	3.5	5.6	3.3	4.3	2.7	1.6	0.6	10	-	24	-	29	89	1946	443	-	1.8	3.9	46	-	-	-	
30	R.K.	21F	1.3	139	82	18.2	3.0	3.8	5.5	4.6	3.7	2.6	1.2	2.1	9	-	25	-	23	63	933	385	-	0.9	3.5	27	-	-	-	
31	K.S.	20F	1.3	188	81	27.3	4.6	6.0	6.4	4.1	3.1	2.9	0.2	1.2	8	-	14	-	12	117	1971	180	-	0.6	6.4	11	-	-	-	
Mean	22	1.5	162	88	20.2	4.0	3.4	5.3	4.1	5.5	3.1	2.3	1.0	7	-	6	-	18	-	20	85	1501	244	-	1.3	4.6	32	-	-	-
s.d.	-	-	16	4	3.8	0.6	1.3	0.6	0.4	1.5	0.4	1.3	0.5	5	-	4	-	4	15	358	111	-	0.4	0.9	11	-	-	-	-	
A.S.D. - Type III.																														
32	K.S.	15F	1.3	142	91	16.0	3.5	3.9	4.1	4.1	3.7	3.5	0.2	0.7	4	-	32	-	53	14	84	1565	199	-	635	1.9	3.2	60	0.7 ²	
33	T.T.	34F	1.4	131	88	17.0	3.5	4.4	4.6	3.8	3.0	2.9	0.2	1.3	5	-	31	-	52	13	75	1478	213	-	755	1.5	3.5	44	0.6 ²	
34	T.N.	14M	1.3	135	87	19.7	3.1	3.8	5.3	4.3	3.6	2.6	1.0	1.8	7	-	30	-	50	17	71	1182	266	-	564	1.8	2.9	63	0.7 ²	
35	S.K.	18M	1.4	124	88	18.5	3.2	3.6	5.3	3.9	3.4	2.4	1.1	1.5	11	-	46	-	75	16	96	1330	257	-	998	2.6	3.3	77	0.6 ²	
36	M.E.	10M	0.9	209	72	13.0	5.5	9.9	9.9	4.0	1.9	1.9	0	2.0	8	-	85	-	137	10	104	1612	205	-	3320	2.5	2.6	92	0.3 ²	
Mean	18	1.3	148	85	21.0	3.8	5.3	6.1	4.0	3.1	2.6	0.5	1.5	7	-	5	-	46	-	73	14	83	1434	228	-	1274	1.9	3.1	63	0.6 ²
s.d.	-	-	64	7	6.6	0.9	2.8	2.5	0.2	0.2	0.5	0.4	0.5	3	-	21	-	33	3	9	500	27	1440	1.1	0.3	11	-	-	0.1 ²	
V.S.D. - Type I.																														
37	A.S.	18M	1.6	169	93	19.9	4.9	2.9	4.9	3.5	5.8	3.5	2.3	0	5	-	13	-	16	90	1384	141	-	0.7	7.0	9	-	-	-	
38	I.K.	19M	1.5	163	97	21.1	3.9	2.7	3.9	4.2	6.0	4.2	1.8	0	4	-	17	-	18	97	1787	173	-	1.1	7.5	15	-	-	-	
39	A.A.	28M	1.6	188	95	21.1	5.3	3.0	5.3	3.4	6.2	3.4	2.8	0	8	-	20	-	21	95	2030	212	-	1.1	7.4	14	-	-	-	
40	M.S.	7M	0.7	160	93	17.2	4.6	2.1	4.6	3.5	7.8	3.5	4.4	0	3	-	13	-	14	85	1880	82	-	1.2	8.5	15	-	-	-	
41	S.K.	29F	1.3	166	94	16.3	4.7	1.2	4.7	3.1	12.5	3.1	9.4	0	8	-	20	-	23	82	1893	115	-	1.8	12.0	15	-	-	-	
42	S.K.	4F	0.6	148	96	15.8	5.0	1.5	5.0	3.5	11.7	3.5	8.2	0	9	-	21	-	23	73	1468	123	-	2.0	9.4	21	-	-	-	
43	T.O.	13F	1.1	154	95	19.0	4.4	1.3	4.4	3.5	12.0	3.5	8.5	0	6	-	35													

described by Dow et al.⁴⁾, is also to be accepted really as a physiological mass.

Thus obtained values of flows, resistances¹⁵⁾ and works were represented in Table 1 as those of per square meter of body surface area in each individual cases, for convenience' sake of direct comparison. "Stenotic resistances", however, were indicated in the absolute values, as they are intrinsically related to the actual stenotic orifice areas.

CLASSIFICATION OF THE SUBJECTS

TABLE I (b) CATHETERIZATION RESULTS OF THE SEVENTY-SEVEN PATIENTS OF CONGENITAL HEART DISEASE (CONTINUED)

Case No.	Name	Age Sex	BSA	O ₂ Cons.	Art. O ₂ Sat.	O ₂ Cap.	A.-V. O ₂ Difference			Blood Flow			Shunt			Mean Pressure					* Vascular Resistance			Ventricular Work			PVMA (DA)
							Sys	Pul	Eff	Sys	Pul	Eff	L-R	R-L	RA	LA	RV	RVa	PA	EA	Sys	Pul	SPVR	R V	L V	R/L	
yr. m ² cc/min/m ² % vol. % lit./min/m ² mm. Hg dynes sec.cm ⁻⁵ kgM/min/m ² % cm ²																											
V.S.D. - Type II.																											
44	Y.M.	11F	1.0	180	87	21.2	4.6	2.7	6.0	3.9	6.6	3.0	3.6	0.9	10	-	39	-	42	88	1597	492	-	3.5	6.8	51	-
45	A.H.	21F	1.3	172	87	16.9	4.6	3.3	6.9	3.7	5.2	2.5	2.7	1.3	2	-	37	-	48	81	1280	504	-	3.2	4.5	71	-
46	H.S.	18M	1.4	161	84	25.8	3.6	2.6	5.3	4.5	6.1	3.0	3.1	1.5	6	-	47	-	74	94	1115	650	-	4.9	6.9	71	-
47	S.U.	16M	1.4	168	81	21.2	4.7	4.6	7.9	3.9	4.0	2.3	1.7	1.6	5	-	45	-	93	95	1350	1290	-	6.9	5.9	117	-
48	E.H.	28M	1.4	190	84	22.6	3.6	4.3	6.7	5.4	4.5	2.9	1.6	2.5	3	-	54	-	81	106	1065	991	-	5.0	5.1	96	-
49	S.H.	23F	1.3	138	71	16.3	3.4	5.2	7.5	4.1	2.7	1.8	0.8	2.3	7	-	42	-	88	95	1321	1901	-	4.9	4.8	104	-
Mean		20	1.3	168	82	20.5	4.1	3.8	6.7	4.0	4.8	2.6	2.2	1.7	6	-	44	-	71	93	1658	1280	-	4.7	5.7	85	-
s.d.		-	-	16	6	3.6	0.6	1.0	0.9	0.3	1.4	0.4	1.0	0.6	3	-	61	-	19	8	216	690	-	1.1	0.9	22	-
V.S.D. - Type III.																											
50	K.Y.	15M	1.3	136	88	18.1	3.6	1.6	5.2	3.7	8.6	2.6	6.0	1.1	7	-	31	52	17	88	1337	92	247	2.8	7.9	35	1.1 ^a
51	M.K.	26M	1.6	131	89	25.2	3.1	2.7	4.7	4.2	4.9	2.6	2.1	1.4	2	-	41	67	16	99	1152	119	531	3.0	6.0	49	0.8 ^a
52	H.O.	22M	1.2	226	85	29.9	8.7	7.9	12.1	2.6	2.9	1.9	1.0	0.7	7	-	49	80	15	125	2810	220	1455	2.0	4.7	43	0.6 ^a
53	K.K.	45M	1.3	197	89	21.5	6.2	8.0	8.3	3.2	2.5	2.5	0.1	0.8	5	-	47	77	20	94	1637	354	1350	1.9	3.4	57	0.5 ^a
54	Y.H.	8M	1.0	154	73	24.4	4.4	5.1	9.0	3.5	3.0	1.7	2.5	2.0	16	11	59	96	20	110	2090	415	2109	1.7	4.3	41	0.4 ^a
Mean		23	1.3	169	85	24.6	5.2	5.1	7.8	3.7	4.4	2.3	2.2	1.3	7	-	45	74	14	103	2501	288	1178	2.6	5.2	45	0.7 ^a
s.d.		-	-	39	7	4.4	2.1	2.1	2.7	0.4	2.4	0.4	2.0	0.7	6	-	9	16	4	13	723	135	630	0.6	1.6	81	0.2 ^a
Fallot - Type I.																											
55	M.K.	12M	1.1	140	88	18.4	3.3	4.0	4.7	4.4	3.7	3.1	0.6	1.3	8	-	41	67	16	70	1010	222	1030	1.5	2.9	52	0.6 ^a
56	K.K.	10F	1.1	160	90	19.2	3.8	5.0	5.3	4.3	3.2	3.0	0.2	1.2	6	-	38	63	14	71	1220	204	1090	1.5	0.9	52	0.5 ^a
57	M.N.	16M	1.4	123	87	19.4	3.5	3.7	5.2	3.5	3.3	2.4	1.0	1.1	3	-	59	96	11	93	1485	104	1500	1.8	3.1	56	0.5 ^a
58	K.S.	24M	1.5	161	76	27.7	4.1	7.9	8.7	3.9	2.1	1.8	0.2	2.1	10	6	38	63	10	93	1132	133	1470	1.7	3.2	54	0.5 ^a
59	I.K.	6M	0.7	173	86	20.5	4.4	4.6	6.2	3.9	3.7	2.8	1.0	1.1	4	-	46	75	9	68	1870	116	1940	1.3	2.6	49	0.4 ^a
60	H.S.	6M	0.7	156	86	18.5	4.3	5.8	6.5	3.6	2.7	2.4	0.3	1.2	6	-	34	56	15	80	2340	409	1700	1.6	2.9	55	0.4 ^a
61	F.A.	6M	0.8	153	86	18.2	4.0	5.0	6.7	3.8	3.1	2.3	0.9	1.5	5	-	48	78	16	75	1820	371	2780	1.9	2.4	77	0.4 ^a
62	K.M.	11M	1.0	170	86	25.3	4.8	7.4	7.5	3.5	2.3	2.3	0.1	1.2	4	-	36	60	12	80	1680	242	1682	1.6	2.4	65	0.3 ^a
Mean		11	1.0	155	86	20.5	4.0	5.4	6.4	3.9	3.0	2.5	0.5	1.4	6	-	43	70	13	79	1511	208	1649	1.6	2.8	58	0.5 ^a
s.d.		-	-	15	6	3.4	0.4	1.5	1.4	0.3	0.6	0.4	0.3	0.1	2	-	8	12	3	9	273	69	510	0.3	0.2	9	0.2 ^a
Fallot - Type II.																											
63	K.A.	9M	1.0	155	77	21.4	4.0	4.9	7.9	3.9	3.2	2.0	1.2	1.9	1	-	39	64	9	79	1952	160	1410	2.4	2.2	107	0.5 ^a
64	S.H.	15M	1.3	136	81	23.2	3.6	4.6	7.1	3.7	3.0	1.9	1.1	1.8	5	-	54	88	12	84	1303	143	1527	2.3	2.5	100	0.5 ^a
65	K.M.	28M	1.4	165	77	23.6	4.3	6.4	8.6	4.2	2.8	2.1	0.7	2.1	3	-	62	100	7	87	1134	43	1950	2.6	2.7	96	0.4 ^a
66	K.T.	13F	1.2	163	81	25.7	4.8	7.7	8.4	3.4	2.1	1.9	0.2	2.1	3	-	35	58	8	75	1400	90	1640	1.7	2.0	85	0.4 ^a
67	E.N.	5F	0.7	140	80	19.3	3.5	4.5	6.4	4.0	3.1	2.2	0.2	1.8	4	-	47	77	11	70	1870	235	2600	2.0	2.1	92	0.4 ^a
68	H.S.	18F	1.4	186	72	26.1	4.1	8.8	9.5	4.5	2.1	2.0	0.2	2.5	3	-	51	83	10	81	990	127	1900	3.0	2.3	133	0.4 ^a
69	E.H.	C.	0.9	162	73	25.7	4.1	8.6	9.7	3.9	1.9	1.7	0.2	2.2	4	-	58	94	9	80	1730	195	4210	2.4	1.8	131	0.3 ^a
70	Y.A.	13F	1.4	150	76	31.8	4.1	9.8	10.0	3.6	1.5	1.5	0.1	2.1	3	-	51	83	9	83	1231	142	2640	2.6	1.7	153	0.4 ^a
71	H.H.	8F	0.8	228	81	32.0	2.7	5.2	5.8	5.4	2.8	2.5	0.9	2.8	3	-	53	86	10	94	1660	174	2690	7.3	3.5	208	0.4 ^a
Mean		13	1.1	167	77	25.3	4.1	6.8	8.0	3.9	2.4	1.9	0.6	2.1	3	-	50	82	10	82	1583	159	2285	2.9	2.4	120	0.4 ^a
s.d.		-	-	14	5	3.9	0.3	1.8	2.1	0.3	0.6	0.7	0.4	0.1	2	-	8	13	2	5	209	59	840	0.4	0.4	27	0.1 ^a
Fallot - Type III.																											
72	E.I.	9F	0.7	158	64	30.8	4.2	7.9	13.6	3.8	2.0	1.4	0.6	2.4	7	-	44	72	12	88	2442	380	3340	2.7	1.4	180	0.4 ^a
73	Y.K.	5M	0.6	167	67	31.5	4.1	10.9	17.5	4.1	1.5	1.0	0.6	2.8	4	-	33	64	7	73	2250	180	5060	2.7	1.3	217	0.2 ^a
74	Y.I.	10M	0.9	165	70	37.5	3.6	11.2	12.0	4.6	1.5	1.4	0.1	3.2	7	11	43	71	13	56	943	473	3433	2.3	1.0	227	0.3 ^a
75	T.K.	7M	0.8	179	47	30.2	4.9	12.4	18.6	3.6	1.5	1.0	0.5	2.6	4	-	41	67	7	80	2247	148	4500	2.9	1.1	278	0.3 ^a
76	S.K.	17M	1.2	142	57	22.7	3.1	11.3	12.3	4.5	1.3	1.2	0.1	3.4	8	-	43	71	12	68	887	381	3070	2.9	1.1	268	0.3 ^a
77	A.O.	12F	1.0	171	67	32.4	3.4	12.2	13.0	4.5	1.1	1.1	0.1	3.4	7	-	63	102	11	70	1171	420	6656	3.2	1.0	235	0.2 ^a
Mean		10	0.8	166	62	31.1	3.7	10.5	14.2	4.3	1.6	1.4	0.4	3.1	6	-	47	74	10	74	1335	275	4327	3.6	1.5	253	0.3 ^a
s.d.		-	-	26	8	4.4	0.7	2.4	4.1	0.4	0.6	0.6	0.3	0.3	2	-	8	13	2	10	261	159	1260	1.7	0.9	36	0.1 ^a

* The vascular resistances in the systemic and pulmonary vasculatures are represented at per square meter of the body surface areas, while the stenotic pulmonary valve resistances (SPVR) are shown in the absolute values.

** PVMA (DA): Stenotic pulmonary valvular mean area (or Patent ductus sectional area measured during surgical operations).

Group B. Atrial Septal Defect (A. S. D.)

- Type I. With interatrial shunt only from left to right. —9 cases.
 Type II. Also with right-to-left one besides the above. —7 cases.
 Type III. Atrial septal defect combined with pulmonary stenosis (Fallot's trilogy). —5 cases.

Group C. Ventricular Septal Defect (V. S. D.)

- Type I. Characterized with normal or sometimes mildly increased pulmonary artery pressure up to 40mmHg. —7 cases.
 Type II. With severe pulmonary hypertension, at times with partial right-to-left shunt (Eisenmenger's complex). —6 cases.
 Type III. Ventricular septal defect combined with pulmonary stenosis (so-called "atypical" Fallot's tetralogy). —5 cases.

Group D. Tetralogy of Fallot (Fallot)

- Type I. Mild cases with arterial oxygen

saturations above 80 per cent.

—8 cases.

Type II. Moderate cases with those of 80–70 per cent. —9 cases.

Type III. Severe cases with those of 70–50 per cent. —6 cases.

RESULTS

(1) Blood Flows in Greater and Lesser Circulations

Among each group of the subjects significant differences were not seen concerning the systemic flows, which were observed within nearly normal ranges, i. e. 2.4–4.4 lit./min./M², in greater part of the cases. Pulmonary flows, however, were remarkably varied with the subjects, depending upon the differences in both amounts and directions of the shunts associated. As the blood flow (lit./min./M²) is to be represented as the product of the oxygen consumption (dl./min./M²) and the reciprocal of the arterio-venous oxygen difference (vol. %) in greater or lesser circulation, as obviously from Fick's principle, both the systemic

Fig. 1. Systemic and pulmonary blood flows in the individual cases of the four grouped subjects are represented at the products of the oxygen consumptions and the reciprocals of the arterio-venous oxygen differences in the systemic and pulmonary circulations. Consequently the same levels of flow volume are ranged on a series of some hyperbolic lines. Normal ranges are indicated by each elliptical boundary.

and the pulmonary blood flows in the individual cases of four groups of the subjects were to be corresponding to the points dotted as in Figure 1, where those of the systemic (black points) remained for the most part within each elliptical boundary, that is equivalent to the limit of normal cardiac index as reported by Kobayashi et al.¹⁶⁾ in Prof. Mikamo Clinic, while those of the pulmonary were apparently deviated from that boundary in many of each four grouped subjects.

The decrease of the effective pulmonary flow, necessarily in case of the apparently reduced pulmonary flow, was remarkable particularly in Fallot's tetralogy. There it was observed that the decrease of the effective pulmonary flow (expressed in percentage for the average of normals, i. e. 3, 6 lit./min./M²) was nearly parallel to the decrease of the arterial oxygen saturation (%) as well as to the increase of the blood hemoglobin content (gr./dl.) calculated from the oxygen capacity (vol. %)⁶⁾, as seen in Figures 2 and 3.

(2) Vascular Resistances Particularly in Pulmonary Circulatory System

According to Poiseuille's law, the vascular resistance is interpreted as the ratio of the pressure fall in a vascular system to the volume of blood flowing at the same time. As in the systemic

circulation both the pressures and the flows were within nearly normal ranges in the greater part of the cases in this paper, there the vascular resistances were found essentially normal, i. e. ca. 1000-2000 dyne se. cm⁻⁵. However, the pulmonary vascular resistances varied largely with the differences in both pressures and flows in the pulmonary circuit, as seen in Figure 4, where the resistance is measured with the angle (gradient) formed by the line through the mark of 5 mmHg on the ordinate (pulmonary pressure head, mmHg) against the abscissa (pulmonary flow, lit./min./M²), providing that the left ventricular diastolic mean pressure remains normal (approximately 5 mmHg)⁵⁾. In the same Figure, the limits of normal resistances are also represented by two solid lines, indicating the appropriate maximum and minimum gradients of the pulmonary arterial pressure to the pulmonary flow within normal limits, which are equivalent to 300 and 50 dynes sec. cm⁻⁵ (per square meter) respectively calculated as absolute values. Thereupon, it seemed apparent that the two different populations were perceived with regard to the points plotted in Figure 4: the one with evidently increased pulmonary resistances, sometimes nearly equal to or somewhat superior to those of the systemic (Fig. 8), but with the relatively restricted pulmonary flows

Fig. 2, Fig. 3. Correlations between the effective pulmonary flow and the arterial oxygen saturation (Fig. 2) as well as the hemoglobin content of the blood (Fig. 3) in the twenty-one cases of the Fallot's tetralogy. Effective pulmonary flow is shown at the per cent of the mean value of the normal pulmonary flow, i. e. 3, 6 lit./min./M². Hemoglobin in grm./dl. is computed dividing the oxygen capacity by the coefficient 1.34 (after Van Slyke).

(3–7 lit./min./M²), consisting of the cases of V. S. D. –II. and P. D. A. –III. types, and the other with nearly normal resistances notwithstanding excessively variable pulmonary flows (1–12 lit./min./M²), consisting of the remaining cases.

Fig. 4. Pulmonary vascular resistances in all of the present cases are indicated by the tangents (slopes) of the pulmonary arterial mean pressures against the pulmonary flows (see text).

(3) *Shunt Volume from Greater to Lesser Circulation (Exemplified by Patent Ductus Arteriosus)*

Quite analogously as the mean orifice area is estimated at the quotient of the mean flow volume divided by the square root of the mean pressure gradient across the orifice as aforementioned in case of the pulmonary stenosis¹³, the mean flow volume (lit./min.) through a direct opening or a short tube (e. g. patent ductus) between the greater and the lesser circulations is inversely to be approximately proportional to the product of the mean orifice area (cm²)—that is in the usual patent ductus regarded to be practically equal to the actual orifice size because of the nearly constant ductus flow—and the square root of the mean pressure gradient across it ($\sqrt{BAm - PAm}$, mmHg)¹⁴. This was examined in the eleven cases of P. D. A. whose ductus areas were all measured during surgical operations, and there it was observed that the actual shunt volume estimated through catheterizations were within the errors less than 1, 0 lit./min. in keeping with the ideal ones, that were the very products of the given orifice areas and the known pressure gradients

and consequently represented by a series of some hyperbolic lines really as in Figure 5, where the plotted points indicating actual shunt volumes were found near by the corresponding hyperbolas.

Fig. 5. In the eleven cases of the operated patent ductus, the ductus areas are plotted against the square root of the aortico-pulmonary pressure gradients, then the corresponding shunt volumes are seen fairly ranged hyperbolicly. The ideal levels of the ductus shunts are indicated actually by a series of hyperbolic lines (see text).

(4) *Some Hemodynamic Data Concerning Pulmonary Stenosis*

In this paper the cases with pulmonary stenosis were observed in three different groups, namely those with atrial septal defect (Fallot's trilogy, as the type III of A. S. D.), with ventricular septal defect (so-called "atypical" tetralogy, as the type III of V. S. D.), and finally the typical tetralogy of Fallot (Fallot-types, I, II, III). In these individual types some hemodynamic peculiarities, for which the pulmonary stenosis is to be most responsible were investigated.

i) In the type III of A. S. D. excepting one case M. K., the pulmonary flows (3, 0–3, 7 lit./min./M²) as well as the pulmonary arterial mean pressures (13–17 mmHg) were in nearly normal ranges, and the right ventricular systolic mean pressures (50–75 mmHg) were only mildly elevated. Thus there was suggested that the pulmonary stenosis was not severe, and indeed the pulmonary orifice areas were calculated practically as $0,6 \times 0,6$ cm² or more, and those stenotic resistances remained within 1000 dyne sec. cm⁻⁵, while in Case M. K. with an apparently reduced pulmonary flow (1, 9 lit./min./

M²) and with a remarkably increased right ventricular pressure (137 mmHg), the pulmonary orifice area was estimated at $0,3 \times 0,3 \text{ cm}^2$ (that was later ascertained fairly identical with the actual orifice size in the autopsy), and the stenotic resistance at $3320 \text{ dyne sec. cm}^{-5}$, that was twice as large as the systemic vascular resistance of the case. The interatrial right-to-left shunt was also most conspicuous in Case M. K., coming up to about a half of the systemic flow, which burdened the left ventricle so much as the right ventricular volume work was lightened on the other, that the ratio of the calculated right to left ventricular work in this case (92%) was placed relatively near by those of the others (up to 72%) notwithstanding the much stricter pulmonary stenosis.

ii) In the typical tetralogy the predominant right-to-left shunt mostly from right ventricle into overriding aorta was characteristic, and thus resultant decrease of the effective pulmonary flow was fairly correlated to the arterial desaturation, as mentioned before. Here the stenotic pulmonary resistances, which were greater than those of the systemic vasculatures almost always in the typical tetralogy, were particularly increased in many cases of the types II and III (ca. $2000\text{--}3000 \text{ dyne sec. cm}^{-5}$) whose pulmonary orifice areas were found

at about $0,3\text{--}0,4 \text{ cm}^2$, while those of the mild cases with pulmonary orifice areas of about $0,4\text{--}0,6 \text{ cm}^2$, many among type-I and somewhat in type-II, remained within the ranges of ca. $1000\text{--}2000 \text{ dyne sec. cm}^{-5}$, that were nearly equal to those of the systemic vasculatures (Figs. 7 and 8). The ratio of the right to left ventricular work was remarkably increased also in the types II and III, coming up to about $100\text{--}300\%$, though in type-I remained within utmost 80%, as indicated in Figure 9. (Further in the same figure, the approximate measures of overriding of aortas in some autopsied cases were schematically indicated, to which some reference will be made later.)

iii) Though similarly consisting of both pulmonary stenosis and ventricular septal defect, the V. S. D.-type III in this paper was believed somewhat differed physiologically from the typical tetralogy, and at times virtually may be called as an "atypical" tetralogy¹⁷⁻¹⁹. Thus there in Cases 50 and 51, at first markedly increased pulmonary flows up to $8,6$ and $4,9 \text{ lit./min./M}^2$ respectively were striking, concurrently with apparent left-to-right intracardiac shunts, disclosing the relatively wide pulmonary orifice areas calculated as $1,1$ and $0,8 \text{ cm}^2$ as well as the mild stenotic resistances of 247 and $531 \text{ dyne sec. cm}^{-5}$ respectively. Similarly

Fig. 6, Fig. 7. Relations between the mean effective area of the stenotic pulmonary orifice (see text) and the pulmonary arterial flow (Fig. 6) as well as the stenotic pulmonary valve resistance (Fig. 7).

in Cases 52 and 53 the pulmonary stenotic resistances were both less than those of the systemic vasculatures, and there also were perceived a predominant left-to-right intracardiac shunt in Case 52 (this case had been clinically quite similar to a isolated pulmonary stenosis), and a somewhat increased pulmonary arterial pressure in Case 53 (a man of 45 years old, clinically doubted of an Eisenmenger's complex). (Thus physiologically, these cases seem to be situated between the typical tetralogy and the isolated ventricular septal defect or even Eisenmenger's complex, and some cases clinically free from apparent cyanosis may be called as an "acyanotic" tetralogy¹⁹⁾, as it was nearly so in Cases 50 and 51.) Finally in Case 54, in which the left atrial blood was shown markedly reduced in the oxygen saturation (75%), a large right-to-left interatrial shunt, calculated up to 2.5 lit./min./M² and over the total left-to-right intracardiac shunt, was most responsible for the increased left ventricular volume work far more than that of the right ventricle, resulting a so much small ratio (41%) of the calculated right to left ventricular work in spite of an apparently reduced arterial oxygen saturation (73%), as compared with those in the cases of typical tetralogy with similar arterial oxygen saturations. (Such a case as above will be called as a "pentalogy" of Fallot, not merely in the usual sense signifying an association of atrial septal defect besides tetralogy but here particularly by a presence of significant right-to-left interatrial shunt causing peculiar hemodynamics, differed from the typical tetralogy and revealed clinically by some electrocardiographic findings among others.)^{5,20)}

DISCUSSION

Part I Pulmonary Circulatory Dynamics in Some Congenital Heart Disease with Special Reference to Pulmonary Vascular Resistance

(1) Some Affairs under a "Pertinent" Pulmonary Vascular Resistance

Though normally pulmonary flow volumes are to be much variable with the size of subjects (usually per square meter of body surface area), pulmonary arterial mean pressures are commonly believed to be not so easily affected by the size and aging of subjects, (normally within the ranges of ca. 13–18 mmHg⁵⁾. Thereupon, the pulmonary vascular resistances (P. V. R.), that are equivalent to the

gradients of pulmonary pressure falls to variable pulmonary flows as aforementioned, will be reasonably standardized, taking account of pulmonary flow volumes per square meter of body surface area¹⁵⁾ (pulmonary flow index, lit./min./M²), as performed in this paper and also indicated in Figure 4. Under such considerations as above, the values of a "pertinent" (somewhat in the meaning of "likely normal") P. V. R. were assumed to be ca. 50–300 dyne sec. cm⁻⁵ (per square meter), through appropriate calculations using normal ranges of the pulmonary arterial mean pressure as well as the pulmonary flow index¹⁶⁾.

Now it will be studied the pulmonary hemodynamics in some congenital heart disease with such a "pertinent" P. V. R., consisting of the greater part of the cases in this paper other than the P. D. A.-III and V. S. D.-II types (Fig. 4). The latter two types were rather characterized with "apparently increased" P. V. R. and will be discussed in the following paragraph.

As one of the most striking facts observed in the cases with pertinent P. V. R. it must be cited that there the pulmonary arterial mean pressures are found within relatively narrow ranges, i. e. ca. 10–30 mmHg for the most part, as compared with the markedly fluctuating pulmonary flows which are now under 2 lit./min./M² in some Fallot's tetralogy and then up to even 12 lit./min./M² in some others. Thus in these cases, there may be deduced that such a "pertinent" P. V. R. as above will act for the pulmonary arterial pressures to be not so much deviated from the normal ranges under exceedingly variable pulmonary flows primarily due to the cardiovascular anomalies, providing that the P. V. R. in general is one of the important regulators of pulmonary hemodynamics, being effectuated through some functional as well as anatomical ways, as it is believed somewhat so for the increased pulmonary flows during exercise at normal persons^{21–23)}. Such adaptive mechanisms of the "pertinent" P. V. R. likely for the pulmonary hemodynamics, however, are at times seemingly insufficient, resulting in some cases somewhat increased pulmonary arterial mean pressures (usually up to 30 mmHg and rarely to about 40 mmHg), which in the author's opinion seem to be rather "functional"²⁴⁾ than "substantial"—chiefly based on some anatomical substrata as discussed later in connection with "apparently increased"

P. V. R.—pulmonary hypertensions. That is to say in these cases, a functional mitral stenosis caused by largely increased pulmonary flows as one of the most probable factors may be responsible for the somewhat increased pulmonary pressures (, though not fully substantiated in this paper, but clinically often preferably deemed so on account of some “apical diastolic murmurs”²⁷). (From a practical point of view, accordingly in the author’s opinion, such hypertensive cases as above with “pertinent” P. V. R. may be much readily improved by some surgical procedures through which increased pulmonary flows are to be normalized, probably as compared with those indicating “apparently increased” P. V. R., not always depending upon the mere differences in the levels of pulmonary arterial pressures.) Sometimes, an actual mitral valve disease may be associated in these above-mentioned cases, as ascertained through catheterization practically in one case of A. S. D. (Case R. T.), whose left atrial pressure was found much elevated to 24 mmHg, here most likely accounting for the increased pulmonary arterial mean pressure up to 37 mmHg. (Recently Lucas et al.^{25,26}) emphasized the “back pressure” effect from the overworked left ventricle as one of the plausible factors responsible for the “functional” pulmonary hypertension as above-mentioned, because the pulmonary flows and necessarily the outputs from the left heart are increased far greater than the normal, such as above 10 lit./min./M² in some cases of congenital heart disease with large left-to-right shunt, as seen also in this paper, thus inviting the increased end-diastolic pressures of the left ventricle.)

(2) Some Consideration with an “Apparently Increased” Pulmonary Vascular Resistance

In the cases with apparently increased P. V. R. above 300 dyne sec. cm⁻⁵ (per square meter) sometimes nearly equal to those of the systemic, mainly consisting of the P. D. A.-III and V. S. D.-II types in this paper, it may be apparent that the pulmonary arterial mean pressures are definitely increased, widely ranging from about 40 to 90 mmHg in many, though the pulmonary flows were relatively confined within ca. 3–7 lit./min./M², and moreover there the pressures seem to be more advanced generally in the cases with the lesser pulmonary flows. Such events as above are to be preferably interpreted something in the manner, that there have been taken place rather some anatomical

changes of pulmonary vasculature exaggerating the actual hindrance for pulmonary flows, which precipitate the rapidly increasing pulmonary arterial pressures and on the other hand bring about some reducing pulmonary flows through lessened pressure gradients between greater and lesser circulations, presumably in some part owing to the more or less reduced capacity of total pulmonary vascular bed associated with vast anatomical degenerations. As some of the anatomical changes just as above assumed, it may be cited the intimal hypertrophy sometimes associated with multiple thrombi in the medium- and small-sized pulmonary arteries, as really observed by many authors^{28–31}) at autopsies of some congenital heart disease with left-to-right shunt, which has been frequently regarded as some “secondary” degenerations following to the abnormally increased pulmonary flow of hitherto long standing (, probably through a state under “pertinent” P. V. R. as mentioned before) and consequently in general may be more easily expected in the relatively older subjects of the cases³²).

However, from another point of view than above, it may be also possible that the pulmonary hypertension has been established primarily at an earlier age of the subject or rather congenitally^{33–41}), so that the pulmonary flow can be retained within certain limits likely as afore-mentioned. It seems particularly so in some cases of V. S. D. or P. D. A. in which the septal defects or the patent ductus are essentially large-sized, for there the left-to-right shunts of extraordinary amounts would not be compatible with the patients’ lives through pulmonary overflows as well as much depleted systemic flows^{24,38}), if the pulmonary arterial pressures were not primarily elevated and so the pressure gradients across the defects or the ductus were not retained small sufficiently for their enough large-sized openings, as properly expected from a before-mentioned fact that the shunt volume through an opening is to be approximately proportional to the size of that opening and as well as the pressure gradient across it (Fig. 5). Indeed in some autopsied cases of the so-called Eisenmenger’s complex, in which the pulmonary arterial pressure is usually nearly equal to that of the systemic under a fairly large-sized ventricular septal defect, Civin and Edwards found a preservation of somewhat fetal characteristics of pulmonary vasculatures³⁵), that was considered as essentially re-

sponsible for the primarily elevated P. V. R. and consequently pulmonary arterial pressures nearly equal to those of the systemic³⁴⁾. Likely somewhat so for certain cases of so-called "wide" patent ductus, as deemed by some authors³⁹⁻⁴¹⁾. In the present series of this paper, a female subject of P. D. A. (Case M. N.) was remarkable for the apparently increased P. V. R. as well as pulmonary arterial pressure nearly equal to those of the systemic already in her three years of age, which seemed to be accountable at times for the partial right-to-left shunt through the ductus when the P. V. R. was more elevated than the systemic, as almost assured by an occasional reduction of arterial oxygen saturation (74%) (and likely from the hitherto clinical evidence of transient cyanosis frequently under emotional states of the subject).

*Part II Hemodynamic Peculiarities under
Some Types of Congenital Pulmonary Stenosis*

(1) Physiological Significance of "Effective" Pulmonary Orifice Area

Since some difficulties are not avoidable for an accurate estimation of actual orifice areas in various types of congenital pulmonary stenosis, as previously mentioned, the "effective" pulmonary orifice areas—rather as physiological measures—are preferably of practical use, reflecting that the mean pulmonary flow (lit./min.) as well as the pressure height of right ventricle (mmHg) are both prescribed through the "mean effective" area (cm²) of any stenosed pulmonary orifice, as appropriately deduced from Bernouille's hydraulic principle¹³⁾.

The pulmonary arterial mean pressures are maintained mostly within 10–15 mmHg even in the severe cases of pulmonary stenosis with much decreased pulmonary flows to about 1.0 lit./min., as afore-mentioned in this paper, probably owing to the regulative mechanism of "pertinent" P. V. R. (or partly through hypoxia with and without polycythemia⁴²⁾ in some cases indicating somewhat increased P. V. R. to about 400 dyne sec. cm⁻⁵)²⁴⁾. So that the values for the "square root" of mean pressure gradients between the right ventricle and the pulmonary artery are not so strikingly varied as those of very pulmonary flows (ca. 1.0–5.0 lit./min., in one case above 8.0 lit./min.), though of course the right ventricular mean pressures are fairly variable among individual cases (ca. 30–60 mmHg, in one case up to 85 mmHg). Thereupon,

from the hydraulic principle just above-cited, it may be deduced that at least for the flow volumes through stenosed pulmonary orifices the "mean effective" area seems to be a main regulative factor, as really indicated in Figure 6. Thus from the same figure, the critical dimensions of that "effective" area with which the pulmonary flow is to be reduced apparently below normal seem to be in approximate values of 0.5–0.4 cm². The most cases of typical tetralogy indicate the "effective" pulmonary orifice areas of the above-cited values or less, while the cases with fairly increased pulmonary flows that are found at times in the V. S. D.-III and A. S. D.-III types of this paper show the "effective" areas of about 0.6 cm² or more.

(2) Some Consideration with Resistance Offered by Stenosed Pulmonary Orifice

The resistance to the ventricular discharge across the stenosed pulmonary orifice (anatomically may be much varied) is called conveniently as "stenotic pulmonary valve resistance" in this paper, and is physiologically estimated in the absolute value of dyne sec. cm⁻⁵, as previously described. (As a rule in this paper, the values of peripheral vascular resistances are expressed at "per square meter" of body surface areas, but obviously must be reversed to the absolute values when necessary that they are placed by the side of the "stenotic" resistances.)

Though in the cases with pulmonary stenosis the pulmonary flows are fairly determined in general by the "effective" pulmonary orifice areas as aforementioned, the distribution of the systemic as well as the pulmonary flow in each individual case seems to be mostly depending upon the individual interrelation of the two resistances to which the both flows are to be opposed, namely the systemic vascular resistance and the "stenotic" pulmonary valve resistance. This seems particularly so in the cases associated with fairly large-sized ventricular septal defect such as typical tetralogy and the type-III of V. S. D., in which the pressure gradients between the left and the right ventricles are considered not so eminent but almost abolished in many^{24,38)}, that the interventricular shunts and in consequence both the systemic and the pulmonary flows seem to be determined almost solely by the individual resistances. Thus, in spite of near the same "orifice areas" of about 0.5–0.6 cm², in some cases of the type-III of V. S. D. and on rare occasions also in

the mildest cases (type-I) of the typical tetralogy, the somewhat reduced "stenotic" as compared to the systemic resistances in the former cases may be favourable for the predominant left-to-right intracardiac shunts, while the reverse relation seems to characterize the right-to-left shunts in the latter, though essentially the two types may be intervening to each other on some occasions (Figs. 7, and 8). (The fairly same circumstances are to be seen in some cases of the type-II of V. S. D.— particularly in the cases of so-called Eisenmenger's complex—, replacing the "stenotic" resistance here by the total pulmonary vascular resistance, which in the very relation to the systemic resistance may be apt to affect the intracardiac shunts rather than the often not so significant pressure gradient between the both ventricles⁴³⁾, as something indicated also in Figure 8.)

The interrelation between the "stenotic" pulmonary resistance and that of the systemic vasculature particularly in the Fallot's tetralogy is to be re-examined in the following paragraph, especially in the connection with the "over-riding" of sorta.

Fig. 8. Comparison of the pulmonary vascular resistance (P. V. R.) or the stenotic pulmonary valve resistance (S. P. V. R.) with the systemic vascular resistance (S. V. R.) in certain types of the subjects. (see the legend in Fig. 9).

(3) Some Particularities from the Aspect of "Ventricular Work"

From a hemodynamic point of view, the work volume of each ventricle is to be approximately represented by the product of individual pressure

height and flow volume with which the both ventricles are to be burdened, and commonly estimated in kg.M./min./M^2 according to the methods as aforementioned¹¹⁾.

As a rule it may be sure that the more the pulmonary stenosis is advanced, the more the right ventricular work is to be increased usually by reason of the more elevated right ventricular pressure. However closely examined, this may be not uniform under the different types of pulmonary stenosis, particularly in the relation to the left ventricular work which will be variably disordered also under the same circumstances, as to be discussed in the following. (Such physiological considerations of the ventricular works may be essentially important for the clinical interpretation of the patients' electrocardiograms, as it will be fully described elsewhere.)²⁰⁾

i) In the type-III of A. S. D. (or so-called trilogity of Fallot) the markedly increased right ventricular pressure due to the advancing pulmonary stenosis seems to cause the so much amounted right-to-left interatrial shunt due to the exaggerated pressure fall from the right to the left auricle, as

Fig. 9. Relation between the pulmonary orific mean area and the ratio of the right to the left ventricular work in the groups combined with pulmonary stenosis.

* Note: The black region in the Aorta indicates the part belonging to the right ventricle, the white to the left; thereby informs the approximate degree of the Aorta-Overriding.

observed previously in Case M. K. of this category, that the right ventricular flow volume tends to be minimized fairly in accordance with the advancing pulmonary stenosis, while that of the left ventricle does not at all fall but occasionally somewhat increased⁴⁴⁾. Thus, in spite of the much increased right ventricular pressures due to the advanced pulmonary stenosis, the ratios of right to left ventricular work seem to be not exceedingly augmented by the reason just above-mentioned but held within a certain limit, seemingly at most 90 per cent as only concerned with the cases of this paper (Fig. 9). (The fairly same affairs as above may be expected in some cases of the so-called pentalogy of Fallot⁵⁾, particularly those with the predominant right-to-left interauricular shunt rather than that of interventricular, as proven in one case of the type-III of V. S. D. or the so-called "atypical" Fallot—that is Case 54 of the series.)

ii) In the tetralogy of Fallot it may be reasonably expected that the ventricular works are to be essentially modified through variable compositions of the trias, namely the pulmonary stenosis, the ventricular septal defect and the "over-riding" aorta, each of which may differ from case to case. Though it must be so in the essential features of the matter, there seems to be deduced some leading principles analytically from the hemodynamic consideration with regard to the ventricular work as follows.

In the usual case of the typical tetralogy that is characterized anatomically with the so-called "high" septal defect as well as "over-riding" aorta participated with both ventricles, the right and the left ventricular pressures are practically to be almost equal, and moreover the intracardiac shunt is regarded for the most part to be taken place directly from right ventricle into "over-riding" aorta, as the amount of left-to-right intracardiac shunt is essentially minute as compared with that of the right-to-left, as really proven in the greater part of the typical tetralogy²⁾. If that is the case, the ratio of the right to left ventricular work, that in this case can be replaced by the ratio of each ventricular flow volume under the same pressure, may be roughly proportional to the ratio of the systemic to the pulmonary flow in the individual cases. Hence the pulmonary flow is to be decreased fairly in accordance with the advancing pulmonary stenosis (Fig. 6), though not so like for the systemic

flow, and then the above-mentioned work ratio may be rapidly increased most probably with the advance of pulmonary stenosis, as really examined in Figure 9, where the exactly calculated ratios of the right to left ventricular work are compared with the "effective" pulmonary orifice areas indicating the various degrees of pulmonary stenosis.

In the same figure, however, it may be not overlooked also that the R/L work ratios seem much variable from about 1/1 to 3/1 in some cases whose "effective" pulmonary orifice areas are of the values in the neighbourhood of 0.3cm^2 , most probably suggesting that in spite of the not so different degrees of pulmonary stenosis the division of the right ventricular discharges towards the aorta and the pulmonary artery may be sometimes much variable and seems to be particularly handicapped for the latter as compared to the former, in some cases with much greater R/L ratios. This may be, preferably only from a physiological point of view, interpretable as that the "stenotic" pulmonary valve resistances in the just-mentioned cases are especially increased far above those of the systemic vasculatures (as somewhat investigated in the preceding paragraph), but rather substantially this seems in most reality to be largely affected by the variable degrees of the "over-riding" of aorta, as properly deemed from the finding in some autopsied cases that the approximate degrees of "dextro-position" of the aortas in these cases are advanced fairly in accordance with the augmentation of the calculated R/L ratios of ventricular works, as schematically indicated also in Figure 9. (As one of the plausible factors accounting for the above observation, it will be suggested that the more the aorta is dextroposed over the right ventricle, the more the right ventricular discharge towards the aorta may be facilitated through the more enlarged outlet under the fairly same right ventricular pressure regardless some variable pulmonary stenosis, while that towards the pulmonary artery, seemingly as a leak—so to speak—from a side aperture in the meaning that the discharging force to the pulmonary stenosis may be determined secondarily depending upon that to the aorta, seems to be somewhat more weakened. In other words, the resistance for the stenosed pulmonary orifice seems the more increased relatively in comparison with that for the enlarging aortic aperture, even if the pulmonary stenosis itself may remain fairly in a same

degree.)

SUMMARY AND CONCLUSIONS

The seventy-seven patients of congenital heart disease consisting of the four groups, namely P.D.A., A.S.D., V.S.D. and Fallot's tetralogy were studied in this paper essentially from a physiological point of view, referring to the several results obtained through catheterization technique, that is the flow volumes including those of various shunts as well as the pressure values in the greater and the lesser circulations, together with the certain well-defined quantities of "vascular resistances", "ventricular works", and especially as for pulmonary stenosis those of "effective pulmonary orifice areas" as well as "stenotic pulmonary valve resistances".

Among the miscellaneous concerning the hemodynamic affairs observed in the present cases of congenital heart disease, special attentions were paid in this paper to (1) the pulmonary hemodynamics particularly in respect to the pulmonary vascular resistance, and (2) the hemodynamic peculiarities concerned with some types of congenital pulmonary stenosis, with which some conclusions were obtained briefly as follows.

(1) Regarding the pulmonary circulation in the various types of congenital heart disease, there were distinguished the two groups mainly with respect to the pulmonary vascular resistance (P.V.R.).

i) The one with "pertinent" P. V. R. that is within ca. 50–300 dyne sec. cm^{-5} (per square meter) was characterized, regardless of the much variable pulmonary flows, with the pulmonary arterial mean pressures of nearly normal levels in many, though at times somewhat increased up to ca. 30–40 mm Hg. seemingly as an expression of functional mitral stenosis caused probably by the exceedingly increased pulmonary flows.

ii) The other with apparently increased P.V.R. above 300 dyne sec. cm^{-5} was distinctive by the remarkably increased pulmonary arterial pressures, though fairly variable and sometimes nearly equal to those of the brachial arteries, against the relatively confined pulmonary flows within ca. 3–7 lit./min./ M^2 .

The plausible genesis as well as the physico-pathological significances of the respective P. V. R. were further discussed.

(2) Detailed hemodynamics were studied throughout various types of congenital pulmonary

stenosis, namely Fallot's trilogy, "atypical" and typical tetralogy of Fallot, specially taking account of some "physiological" measures such as "effective pulmonary orifice area" as well as "stenotic pulmonary valve resistance" which were both in this paper well-defined through appropriate estimations respectively.

i) The mean flow volumes (lit./min.) through any stenosed pulmonary orifice were regarded to be fairly dependent upon the "mean effective pulmonary orifice areas", which critically under the values of ca. 0.4–0.5 cm^2 seemed to bring about the pulmonary flows that were reduced apparently less than normal.

ii) Besides, from another point of view, the respective discharges of the pulmonary and the systemic flows in each individual cases were most likely determined through each interrelation between the "stenotic" pulmonary resistance and that of the systemic vasculature, resulting a predominant intracardiac shunt of either left-to-right or right-to-left, particularly in the cases combined with large ventricular septal defect. That is to say, in the case of the so-called "atypical" Fallot, of which the pulmonary flows were often fairly increased, there seemed the "stenotic" resistances under the levels of those of the systemic in general to be responsible for the predominant left-to-right shunts, just as reversely in the most cases of the typical tetralogy.

iii) Furthermore, the minute differentiation of the typical tetralogy from the "atypical" as well as the "trilogy" of Fallot was investigated from the aspect of the physiologically estimated ratios of the right to left ventricular works, of course in close relation to the variable degrees of pulmonary stenosis and as well of the "over-riding" of aorta, which were in this paper particularly discussed with respect to some cases of typical tetralogy of Fallot.

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