EXPERIENCE WITH CONGENITAL HEART DISEASE AT GROOTE SCHUUR HOSPITAL, CAPE TOWN

AN ANALYSIS OF 1,439 PATIENTS STUDIED OVER AN ELEVEN-YEAR PERIOD

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During the 11-year period 1952 to 1962, Groote Schuur Hospital has been the main hospital serving the population of Cape Town and the surrounding districts. The two main population groups from whom the patient material is drawn are the Whites and the Cape Coloured. As previously analysed,[1,2] the hospital attendance and admission rate is approximately the same for these two racial groups, and so is the population distribution. The Bantu, however, form only 10% of the population and are a far more unstable and unrepresentative group. Comparison of diseases occurring in the three racial groups is therefore far more valid between the Whites and Coloured than for the Bantu.

Groote Schuur Hospital is a general hospital admitting patients of all ages. However, a neonatal service has not been available until very recently and with the establishment of the Red Cross War Memorial Hospital for children in June 1956 much of the paediatric material has been diverted. Diseases with a high mortality and morbidity in early infancy are therefore poorly represented in the hospital statistics. Moreover, with the development of cardiac surgery during the past 4 years, more patients with operable heart disease have been referred, so that these conditions are over-represented.

Despite the fact that severe congenital heart malformations incompatible with long survival are under-represented, and operable conditions over-represented, it was thought worth while to analyse experience in congenital heart disease at the Cardiac Clinic during the past 11 years. As has recently again been emphasized[3] there is at present no way of determining the exact incidence of cardiac malformations in the general population at birth. Many pathological studies have been made but these have perforce been conducted on a highly selected population group. The findings in this survey have therefore been discussed in relation to comparable clinical studies performed in Great Britain,[4] the United States,[5,6] Canada,[7] Sweden,[8] and Australia.[9]

Material and Methods

All patients suffering from congenital heart disease seen in the Cardiac Clinic, Groote Schuur Hospital, in the 11 years 1952 - 62 inclusive have been included in this analysis. In the early years less than 200 new patients in all attended annually but, with the growth of the clinic, that number has grown to over 1,100. Congenital heart disease contributes 20% of the patients examined in this clinic. *Pari passu* with the increase in number of patients attending has been the increased accuracy of clinical diagnosis, the development and expansion of diagnostic techniques, and the progress in cardiac surgery. Patients seen at the Red Cross War Memorial Hospital have not been included in this analysis.

The diagnosis has been based on the clinical examination, electrocardiogram (ECG), and X-ray (fluoroscopic or radiological) findings in every patient. With few exceptions I have personally examined every subject, often on several occasions. In 804 patients cardiac catheterization and/or angiocardiography has been performed in our laboratory (the techniques have been described elsewhere[10]). Surgical treatment has been applied to 540 patients and necropsy study has been carried out in 21 cases. Confirmation of the clinical diagnosis has thus been obtained in at least two-thirds of the patients.

Congenital heart disease was recognized in 1,439 patients (Table I). By far the commonest reason for referral was

TABLE I.* DIAGNOSIS, SEX AND RACE IN 1,439 PATIENTS WITH CONGENITAL HEART DISEASE

	M	*F*	*EM*	*EF*	*CM*	*CF*	*BM*	*BF*	*Total*
Ventricular septal defect	154	159	86	97	61	55	7	7	313
Atrial septal defect	95	150	54	96	34	48	7	6	245
Patent ductus arteriosus	73	159	32	88	34	61	7	10	232
Tetralogy of Fallot	107	68	62	39	38	27	7	2	175
Pulmonary stenosis	64	69	36	51	24	16	4	2	133
Coarctation of aorta	56	32	43	23	11	9	2	0	88
Aortic stenosis	40	24	31	17	9	6	0	1	64
Miscellaneous	93	96	44	63	37	32	12	1	189
Totals	682	757	388	474	248	254	46	29	1,439
			862		502		75		

* Throughout Tables I-X: E (European) = White. C = Coloured. B = Bantu. M = male. F = female.

the presence of a cardiac murmur. Less commonly attention was drawn to the heart because of symptoms, cyanosis, failure to thrive, or abnormal ECG or X-ray findings. The incidence of individual forms of congenital heart disease according to sex and race is shown in Table I. The sex, race and age incidence is shown in Table II. Where multiple defects were present, the major anomaly was assessed and the case included only in that group.

TABLE II. SEX, RACE AND AGE IN 1,439 PATIENTS WITH CONGENITAL HEART DISEASE

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	364	361	198	222	138	124	28	15	725
10–19	171	196	94	120	68	67	9	9	367
20–29	69	101	39	56	26	41	4	4	170
30–39	43	46	32	34	8	11	3	1	89
40–49	27	31	19	25	6	6	2	0	58
50+	8	22	6	17	2	5	0	0	30
Totals	682	757	388	474	248	254	46	29	1,439

1. *Ventricular septal defect.* Of the 313 patients, the diagnosis was confirmed by cardiac catheterization in 185 and surgical correction was attempted in 43. At least 20 patients had associated defects such as atrial septal defects, coarctation of the aorta, and pulmonary stenosis. Patent ductus arteriosus was a common complication sometimes found at operation and unsuspected clinically. The sex, race and age incidence is shown in Table III.

TABLE III. SEX, RACE AND AGE IN 313 PATIENTS WITH VENTRICULAR SEPTAL DEFECT

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	101	92	54	57	41	30	6	5	193
10–19	35	41	19	23	16	16	0	2	76
20–29	10	13	6	9	3	4	1	0	23
30–39	6	6	5	3	1	3	0	0	12
40–49	1	6	1	4	0	2	0	0	7
50–59	1	1	1	1	0	0	0	0	2
Total	154	159	86	97	61	55	7	7	313

2. *Atrial septal defect.* Of the 245 patients, the diagnosis was confirmed by catheterization in 170 and surgical correction was attempted in 96. One-fifth of the patients had endocardial-cushion defects. Pulmonary stenosis was associated in 18. The sex, race and age incidence is shown in Table IV.

TABLE IV. SEX, RACE AND AGE IN 245 PATIENTS WITH ATRIAL SEPTAL DEFECT

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	24	52	7	37	14	13	3	2	76
10–19	26	34	17	14	7	17	2	3	60
20–29	13	31	6	19	6	11	1	1	44
30–39	14	13	12	11	1	2	1	0	27
40–49	14	10	10	7	4	3	0	0	24
50–59	2	6	1	6	1	0	0	0	8
60+	2	4	1	2	1	2	0	0	6
Total	95	150	54	96	34	48	7	6	245

3. *Patent ductus arteriosus.* Of the 232 patients, operation was performed in 162. 16 patients were catheterized. Subacute bacterial endocarditis occurred in 11. The sex, race and age incidence is shown in Table V.

TABLE V. SEX, RACE AND AGE IN 232 PATIENTS WITH PATENT DUCTUS ARTERIOSUS

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	43	75	18	39	21	32	4	4	118
10–19	14	42	4	24	10	15	0	3	56
20–29	7	20	4	8	3	9	0	3	27
30–39	5	9	4	7	0	2	1	0	14
40–49	4	6	2	5	0	1	2	0	10
50–59	0	5	0	3	0	2	0	0	5
60–69	0	2	0	2	0	0	0	0	2
Total	73	159	32	88	34	61	7	10	232

4. *Tetralogy of Fallot.* Of the 175 patients the diagnosis was confirmed by catheterization or angiocardiography in 145 and by operation or necropsy in another 9. Operation has been performed on 86 patients, 65 by the cardiac bypass and open-heart procedure in our institute. The sex, race and age incidence is shown in Table VI.

TABLE VI. SEX, RACE AND AGE IN 175 PATIENTS WITH TETRALOGY OF FALLOT

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	74	47	46	24	23	21	5	2	121
10–19	27	10	13	8	14	2	0	0	37
20–29	5	6	2	3	1	3	2	0	11
30–39	1	4	1	3	0	1	0	0	5
40–49	0	1	0	1	0	0	0	0	1
Total	107	68	62	39	38	27	7	2	175

5. *Isolated pulmonary stenosis.* Of the 133 patients, cardiac catheterization was performed in 105 and operation in 53. The sex, race and age incidence is shown in Table VII.

TABLE VII. SEX, RACE AND AGE IN 133 PATIENTS WITH PULMONARY STENOSIS

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	31	24	20	20	9	3	2	1	55
10–19	20	26	11	21	7	5	2	0	46
20–29	8	12	3	6	5	6	0	0	20
30–39	4	6	2	3	2	2	0	1	10
40–49	1	1	0	1	1	0	0	0	2
Total	64	69	36	51	24	16	4	2	133

6. *Coarctation of the aorta.* Operation was performed in 53 of the 88 patients. Complications such as ventricular septal defect and aortic valve disease were present in 10 and tetralogy of Fallot in 1. Patent ductus arteriosus was not uncommon, usually with postductal coarctation.

TABLE VIII. SEX, RACE AND AGE IN 88 PATIENTS WITH COARCTATION OF AORTA

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	21	11	17	9	3	2	1	0	32
10–19	14	9	9	5	4	4	1	0	23
20–29	12	7	9	4	3	3	0	0	19
30–39	3	1	3	1	0	0	0	0	4
40–49	6	2	5	2	1	0	0	0	8
50–59	0	2	0	2	0	0	0	0	2
Total	56	32	43	23	11	9	2	0	88

There were 2 patients with coarctation of the abdominal aorta, and 4 with subacute bacterial endocarditis. Patients with 'pulseless disease' or aortic arteritis have been excluded. The sex, race and age incidence is shown in Table VIII.

7. *Aortic stenosis.* The sex, race and age incidence in the 64 patients is shown in Table IX. Catheterization was performed in 22 and operation in 7.

TABLE IX. SEX, RACE AND AGE IN 64 PATIENTS WITH AORTIC STENOSIS

Age	M	F	EM	EF	CM	CF	BM	BF	Total
0–9	13	11	11	6	2	4	0	1	24
10–19	18	7	13	5	5	2	0	0	25
20–29	5	2	4	2	1	0	0	0	7
30–39	4	2	3	2	1	0	0	0	6
40–49	0	2	0	2	0	0	0	0	2
Total	40	24	31	17	9	6	0	1	64

8. *Miscellaneous conditions.* The sex, race and age in 189 remaining patients classified as suffering from 'miscel-

TABLE X. DIAGNOSIS, SEX AND RACE IN 189 PATIENTS WITH MISCELLANEOUS FORMS OF CONGENITAL HEART DISEASE

	M	*F*	*EM*	*FF*	*CM*	*CF*	*BM*	*BF*	*Total*
Tricuspid atresia ..	11	9	5	5	3	4	3	0	20
Transposition ..	19	7	11	3	7	4	1	0	26
Pulmonary atresia	9	8	6	8	2	0	1	0	17
Ebstein's anomaly	7	9	3	6	2	3	2	0	16
Dextrocardia ..	5	7	1	5	2	2	2	0	12
PA Dil. & PI ..	9	15	6	8	3	7	0	0	24
Heart block ..	2	3	2	2	0	1	0	0	5
Fistulae	6	5	3	3	3	1	0	1	11
Etc.*	25	33	7	23	15	10	3	0	58
Totals ..	93	96	44	63	37	32	12	1	189
	189		107		69		13		

* Anomalous pulmonary venous drainage 7. Aortic window 2. Obstructive cardiomyopathy 4. Truncus, common ventricle 10. Right bundle-branch block and murmurs 3. Valve disease 6. Abnormal vessels 4. Sinus of Valsalva 3. Unknown 19. (Total 58.)

laneous forms of congenital heart disease' is shown in Table X. The diagnosis was confirmed by catheterization, operation or necropsy in 106.

ANALYSIS OF THE DATA

Congenital heart disease is a common condition. It accounts for 7% of all patients referred to this hospital for ECG investigation and 20% of all patients referred to the Cardiac Clinic. Since Groote Schuur Hospital is a general hospital for patients of all ages, these figures are naturally far smaller than in a paediatric institution such as the Red Cross War Memorial Children's Hospital.[11]

Congenital heart disease is said to be commoner in Whites than in Negroes,[12] and this is borne out when a comparison is made between the Whites and Coloured in this series (862:502—Table I), confirming previous findings.[1] As stated above, the population at risk, the attendance at the general outpatient department of the hospital, and the admission rate to hospital, are approximately equal for Whites and Coloured, so that the figures probably have significance. No conclusions can be drawn about the Bantu since the true population at risk is unknown. No sex difference was noted in this series. Published necropsy data suggest a greater incidence of congenital heart disease in males,[3] but this has not been the general clinical experience.[7]

The three commonest isolated conditions found were ventricular septal defect, atrial septal defect and patent ductus arteriosus, followed by tetralogy of Fallot, pulmonary stenosis, coarctation of the aorta, and aortic stenosis.

TABLE XI. INCIDENCE OF MAJOR CONGENITAL CARDIAC LESIONS IN PATIENTS WITH CONGENITAL HEART DISEASE: SELECTED CLINICAL STUDIES

	Wood[4]	*Keith* et al.[7]	*Stuckey*[8]	*Nadas*[6]	*Blumenthal*[5]	*Schrire*
Number of patients	690	?	426	577	350	1,439
	%	%	%	%	%	%
Ventricular septal defect	14·5	22	20	11·8	17·7	22
Atrial septal defect ..	23	7	11	16·8	16·6	17
Patent ductus arteriosus ..	13	17	18	17·5	12	16
Tetralogy of Fallot ..	14	11	15	9·4	18·3	12
Pulmonary stenosis ..	11·6	7	8·6	14·4	11	9
Coarctation of aorta ..	6	6	3·7	11	5·7	6

The age constitution of the case groups in this table is as follows: Infants and children: Keith *et al.*, Stuckey and Nadas. All ages, mostly children and adults: Wood and Blumenthal. All ages: Schrire.

As is only to be expected, at least 50% of the patients were recognized before the age of 10; 6% were over the age of 40. Comparison with 5 selected series from Great Britain, Canada, the United States and Australia is shown in Table XI.

1. *Ventricular Septal Defect*

The clinical findings and diagnosis of this condition have been discussed elsewhere.[13] In brief, 3 groups are recognized. In the first, normal right ventricular and pulmonary arterial pressures are present, the defect being minute or small, with a loud parasternal murmur as the only abnormality. The diagnosis is generally easy; amyl-nitrite inhalation or phenylephrine injection[14,15] are often of great help in differentiating this condition from pulmonary or aortic stenosis, innocent systolic murmurs, pulmonary ejection murmurs, or tricuspid valve disease. Cardiac catheterization is not generally necessary for establishing the diagnosis. In the second group, moderate right ventricular and pulmonary arterial hypertension are present. In addition to the parasternal murmur, signs of right ventricular overload and a mitral diastolic murmur are present. In the third, the systemic and pulmonary systolic arterial pressures are similar. The murmur is shorter and softer and the signs of pulmonary hypertension dominate the picture. Where the pulmonary resistance exceeds the systemic, cyanosis without murmurs is present. Cardiac catheterization was performed in almost all patients in the second or third group.

Isolated ventricular septal defect was the commonest single lesion in the White and Cape Coloured, accounting for 22% of the patients with congenital heart disease. The maximum incidence was in the first 2 decades of life, and after the age of 40 it was rare. This may well be due to spontaneous closure of the defect, the true incidence of which has yet to be ascertained.[18] There was an equal incidence in the two sexes in all races; this has been the general experience.[4,8,16,17] Keith *et al.*[7] have pointed out how difficult it is to estimate accurately the true frequency of ventricular septal defect. Necropsy evidence is clearly inadequate, not only because of case selection but because it excludes patients in whom spontaneous closure[18,19] occurs.

The majority of cases of ventricular septal defect are in children under 10 (62% in our series—see Table III), so that in series that do not include infants the reported incidence is relatively low.[4] Our finding that ventricular septal defect constituted 22% of our cases of congenital heart disease tallies with the 22% reported by Keith *et al.*[7] (Table XI).

Ventricular septal defect is frequently associated with other lesions, the commonest of which is pulmonary stenosis. Thus there were 175 patients in which it was associated with severe pulmonary stenosis (tetralogy of Fallot), and 37 with moderate pulmonary stenosis. Other associations are atrial septal defect, coarctation of the aorta and patent ductus arteriosus. A ventricular septal defect is an integral component of the complete variety of endocardial-cushion defect[20] and of many of the complex anomalies such as truncus and pulmonary atresia. If all the conditions are included in which ventricular septal defects were present then ventricular septal defects were found in approximately 40% of all patients with congeni-

tal heart disease. It was found in 34% of the series reported by Keith *et al.*[7]

2. *Atrial Septal Defect*

The clinical findings and diagnosis have been discussed elsewhere.[20,21] Endocardial-cushion defects have been included with secundum and sinus venosus defects. One patient with anomalous pulmonary venous drainage without an atrial septal defect and 3 patients with cor triatriatum have been included. Diagnosis rarely gave rise to difficulty except in small defects, where cardiac catheterization was necessary.

Isolated atrial septal defect (constituting 17% of our cases of congenital heart disease) and patent ductus arteriosus (constituting 16%) were the two commonest conditions after ventricular septal defect, the incidence being approximately the same in the three racial groups. The maximum incidence of atrial septal defect was in the first 3 decades of life, endocardial-cushion defects having a worse prognosis than secundum defects.[20] There were 38 patients over the age of 40 (16%), the oldest in the series being 68. A high incidence of atrial septal defect has been recorded in all series that comprise a large number of adults,[22,23] and the defect is compatible with a long life span—in fact it is estimated that 50% live more than 40 years.[3] Nearly one-third of the 347 adult patients in this series (over the age of 20) had atrial septal defects, but only one-eighth of those under the age of 20. A sex incidence of 3 females to 2 males is in keeping with general experience.[21]

Atrial septal defects are frequently associated with other lesions, apart from endocardial-cushion defects, which of course almost always have atrioventricular valve deformities and often ventricular septal defects. Thus ventricular septal defect, pulmonary stenosis and Fallot's tetralogy were found complicated by atrial septal defect. An atrial septal defect is an integral component of tricuspid atresia and total anomalous pulmonary venous drainage.

3. *Patent Ductus Arteriosus*

The clinical findings and diagnosis have been discussed elsewhere.[13] In brief, 3 groups are recognized. In the first —and by far the commonest—normal pulmonary-artery pressures are present, associated with a typical continuous murmur in the first and second left intercostal spaces. The auscultatory diagnosis was found to be in error in only 2 of over 150 surgically treated patients; one of them had an aorticopulmonary window and the other ventricular septal defect with aortic incompetence. In the second group, significant pulmonary hypertension is present and diagnostic difficulties arise mainly in young infants. In the third, severe pulmonary hypertension coexists. In both the second and third groups the murmur is often atypical, being systolic only, or even absent. Continuous murmurs were found when there were communications between aorta and pulmonary artery (patent ductus arteriosus, aorticopulmonary window, and fistulae between bronchial and pulmonary artery), communications between systemic artery and vein (intercostal, coronary, or mammary souffle), aortic communications with vena cava or any cardiac chamber, rupture of sinus of Valsalva, coronary arteriovenous fistulae,[24] syphilitic aneurysm,[25] penetrating wounds,[26] and pulmonary arterio-venous fistulae.[27] Similar murmurs were encountered in venous hums, total anomalous pulmonary venous drainage, coarctation of the aorta, and distal pulmonary stenosis.[28] Occasionally ventricular septal defect with aortic incompetence produced continuous murmurs.[29,30] Whenever the murmur is atypical either in character or in situation cardiac catheterization should be performed before operation is advised.

As an isolated defect patent ductus arteriosus was found in 16% of patients, being approximately the same in all three racial groups. The maximum incidence was in the first two decades of life, half the patients appearing before the age of 10. The incidence found among paediatric groups by other workers was noted to be 17 - 18%[6,7] and in groups of all ages 12 - 13%[4,5] (Table XI). A sex incidence of approximately 3 females to 2 males was found in all three racial groups. A 2 - 3: 1 female to male incidence has generally been noted.[7,8,30-32]

Patent ductus arteriosus was frequently associated with other congenital cardiac lesions such as ventricular septal defect, coarctation of the aorta, and pulmonary atresia. It was found once only in pulmonary stenosis, Fallot's tetralogy and Ebstein's anomaly.

4. *Tetralogy of Fallot*

This is the commonest cause of cyanotic heart disease, especially after the first year of life. In this series it accounted for about 70% of the patients with cyanosis. The clinical findings and differential diagnosis, with particular regard to the use of amyl nitrite and phenylephrine, have been reported elsewhere.[13,33-37] Tetralogy of Fallot occurred in 12% of the 1,439 patients with congenital heart disease, the ratio being about the same in each racial group. Comparison with findings elsewhere is shown in Table XI. The maximum incidence was in the first decade of life. The oldest patient was 45. 60% of the White and Coloured patients were males, which is the usual proportion noted[7] (Table VI).

Associated defects were not common. Isolated patients had patent ductus arteriosus, coarctation of the aorta, absent pulmonary valves, dextrocardia, and absent left pulmonary artery. The commonest association was atrial septal defect.

5. *Isolated Pulmonary Stenosis*

The diagnosis of pulmonary stenosis with intact ventricular septum has been fully discussed elsewhere, 4 types being noted, namely valvar, infundibular, supravalvar, and distal pulmonary-artery stenosis.[13] Most of the patients had valvar stenosis. No consistent sex difference was found in the three racial groups, and an equal sex incidence was noted in the group as a whole (Table VII). No sex difference has been noted by other authors.[7,38]

The incidence of this condition was 9% — about the same in the three racial groups. The maximum incidence was in the first three decades of life (Table VII) the oldest patient being 46 years.

Pulmonary stenosis is frequently associated with other defects, most commonly ventricular septal defect and atrial septal defect. Almost any other anomaly may be complicated by pulmonary stenosis. There were at least 230

patients with associated pulmonary stenosis, making a total incidence of 25% of the 1,439 patients.

6. *Coarctation of the Aorta*

Except in infants, the diagnosis is readily made by routine palpation of the femoral arteries and the frequent findings of systemic hypertension in youth. It is convenient to separate coarctation in patients over a year from coarctation under a year.[13] Most of the patients (63 out of 88) fell into the former group. As in other series (Table XI) there was a 6% incidence of this condition. Most patients were found in the first three decades of life, and males outnumbered females by almost 2:1 (Table IX), as in other series.

The commonest associated defects were patent ductus arteriosus, aortic valve disease (bicuspid valves), and ventricular septal defect.

7. *Aortic Stenosis*

There were 57 patients with aortic valve stenosis, 6 with subaortic stenosis, and 1 with supra-aortic stenosis. In the advanced case the diagnosis was easy, because of the characteristic murmur, anacrotic pulse, and left ventricular hypertrophy. In the mild case the diagnosis is far more subject to error. Important clues are the radiation of the murmur to the right of the sternum and into the vessels of the neck, the presence of an aortic ejection click, and the accentuation of the murmur on amyl-nitrite inhalation. Long-term follow-up was frequently necessary before making the diagnosis, since left heart catheterization was not felt to be justified in such patients. In patients over the age of 20, a history of a murmur since early childhood in the absence of rheumatic fever was regarded as congenital. It is very probable that many cases of congenital aortic stenosis have been regarded as acquired because of the initial mildness of the lesion, so that probably the high incidence given for the first two decades of life is exaggerated. However, an incidence of 4·4% (Table I) is probably reasonably accurate. In the series reported by Keith *et al.*[7] 4% were due to aortic stenosis, the male to female incidence being 3:2. A similar sex incidence was found in Cape Town and elsewhere.[7,8]

The commonest association was coarctation of the aorta. Obstructive cardiomyopathy producing aortic stenosis was present in 2 patients.

8. *Miscellaneous Conditions* (See Table X)

The diagnosis of *tricuspid atresia* was readily made in a cyanotic child showing left axis deviation in the ECG. Of the 20 children with this condition, 18 were under 10, and there was no sex difference. The diagnosis was confirmed by catheterization or necropsy in 10 of them. Pulmonary oligaemia was far more commonly encountered than normal or plethoric lungs.

Transposition of the great vessels was found in 26 patients, 21 of whom were under 10. The oldest was 28. Males were far more commonly affected than females (3:1), which is the usual experience.[7] Because of the few infants seen in Groote Schuur Hospital, transposition has not been sufficiently represented in this series. In 21 cases the diagnosis was made by catheterization, or at operation or necropsy. Corrected transposition was encountered in 6 patients[28] not included in the transposition group.

Pulmonary atresia was differentiated from Fallot's tetralogy by the presence of continuous murmurs and an aortic ejection click.[13] Confirmation by angiocardiography or necropsy was obtained in 13 of the 17 cases. The sex distribution was equal. Pulmonary atresia with intact ventricular septum has been encountered in 5 patients at the Red Cross War Memorial Children's Hospital,[29] not included in this analysis.

Ebstein's anomaly can be recognized by the characteristic clinical, ECG, and X-ray findings described elsewhere.[13] Seven of the 16 patients were under 16, the oldest being 36. There was an equal sex incidence. The diagnosis was confirmed by catheterization in 12, operation in 2, and necropsy in 1. Four patients with isolated tricuspid valve disease were included in this group.

Dextrocardia was noted in 12 patients and complicated defects were present in 9 of them.

Idiopathic pulmonary-artery dilatation and/or *pulmonary incompetence* may be diagnosed on auscultation,[40] but is generally discovered on X-ray examination. Cardiac catheterization is sometimes required to differentiate the condition from pulmonary stenosis. The age incidence ranged from 6 to 65. 15 of the 24 patients were females.

Isolated complete or partial heart block was found in 5 patients. Complete block complicated one case of ventricular septal defect and one of endocardial-cushion defect.

Arteriovenous fistulae of congenital origin were found in 11 patients, 4 involving the pulmonary artery.

The remaining 58 patients are detailed in Table X. Cardiac catheterization and angiocardiography were generally required for diagnosis, and some cases were only diagnosed at necropsy.

SUMMARY

1. The 1,439 patients suffering from congenital heart disease seen in the Cardiac Clinic, Groote Schuur Hospital, Cape Town, during the 11 years 1952 - 62 are analysed. The diagnosis was confirmed by cardiac catheterization, operation or necropsy in at least two-thirds of the cases and was made on good clinical grounds in the remainder.

2. The incidence of all conditions was higher in the Whites than in the Cape Coloured. The Bantu were too few for analysis and, unlike the Cape Coloured, the Bantu population at risk is too little known for drawing conclusions.

3. Isolated ventricular septal defect was the commonest lesion met with, accounting for 22% of the patients. Combined with other lesions it was present in 40%. Atrial septal defect accounted for 17%, patent ductus for 16%, tetralogy of Fallot for 12%, and isolated pulmonary stenosis for 9%. Pulmonary stenosis associated with other lesions was found in 25%. Coarctation of the aorta and aortic stenosis accounted for approximately 6% each. The remainder consisted of a miscellaneous group of conditions. The incidence of each condition in Whites and Cape Coloured was about the same.

4. The age and sex incidence is analysed in each condition and compared with clinical reports from Great

Britain, the United States, Canada and Sweden. A fairly close agreement is noted.

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