Congenital Heart Disease at Necropsy in Uganda

A 16-years Survey at Mulago Hospital, Kampala

J. B. WOOD, J. SERUMAGA, AND M. G. LEWIS

From the Departments of Medicine and Pathology, Mulago Hospital, Kampala, Uganda

Little is known about the incidence or pattern of congenital heart disease in tropical Africa, except for the description of 15 cases found among 4220 newborn children in Ibadan, Nigeria (Gupta and Antia, 1967), and a report of 44 patients who attended a paediatric cardiology clinic in Kampala, Uganda (Caddell and Connor, 1966). The purpose of this paper is to describe the cases of congenital heart disease found at necropsy in the past 16 years at Mulago Hospital, Kampala.

SUBJECTS AND METHODS

The records from January 1951 to March 1968 were examined, and the cardiovascular abnormalities, age, sex, tribe, cause of death, and congenital abnormalities were listed. Some cases had multiple abnormalities, and these have been listed under that which seemed most important. Simple "probe-patency" of the foramen ovale has been ignored, and patent ductus arteriosus under the age of 1 month has only been recorded when it was wide or associated with other cardiac abnormalities.

The tribes of all cases of congenital heart disease were recorded except for one adult who was admitted unconscious. For comparison, the tribes were recorded of 726 children aged 12 months or less who had necropsies in the years 1951 to 1959 and 1961 to 1966 inclusive. The largest number of cases were Ganda, the people who live and farm in Buganda, a rich, fertile part of Uganda around Kampala. A large local minority comes from the neighbouring countries of Rwanda and Burundi, often to work on the farms—these are listed together as Rwandan. Smaller local minorities come from distant parts of Uganda (e.g. Ankole and Toro) or Kenya (e.g. Luo) to work in Kampala itself.

RESULTS

Among 9432 post-mortem records 60 cases of congenital heart disease were found (6.4 per 1000). There were 32 male cases (53%), 27 female cases, and 1 child with such severe abnormalities that its

sex could not be determined. Thirty-eight of the cases (63%) were aged 12 months or less, and were found among 726 post-mortem records in this age-group (5.25 per 100). The cardiac abnormalities are listed in Table I and described in more detail

TABLE I

TYPES OF CONGENITAL HEART DISEASE FOUND AT NECROPSY BETWEEN JANUARY 1951 AND MARCH 1968

Type	No.	Sex		Associated Abnormality
		M	F	
Atrial septal defect	12	6	6	5
Atrial septal defect and mitral stenosis	2	1	1	—
Atrial septal defect and pulmonary stenosis	1	—	1	—
Atrial septal defect and anomalous pulmonary drainage	2	1	1	—
Ventricular septal defect	9	6	3	1
Single ventricle	3	1	2	1
Ventricular septal defect and overriding aorta	3	—	3	1
Tetralogy of Fallot	2	1	1	1
Valve atresia	6	3	3	1
Patent ductus arteriosus alone	4	1	3	—
Coarctation of the aorta alone	1	1	—	—
Patent ductus arteriosus and coarctation	3	2	1	1
Patent ductus arteriosus and coarctation, and atrial septal defect	1	1	—	—
Valve cusp abnormality	5	5	—	—
Situs inversus	2	1	1	—
Fibro-elastosis	2	2	—	—
Subaortic stenosis	1	—	1	—
Persistent left superior vena cava	1	?	?	1
Total	60	32	27	12

below.* Atrial septal defect was the most common abnormality, and ventricular septal defect was the second most common one.

Abnormalities in other systems were found in 12 cases (20%) of congenital heart disease (Table II).

* A fuller summary of the protocols of the patients can be obtained from J.B.W.

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TABLE II

OTHER CONGENITAL ABNORMALITIES FOUND IN PATIENTS WITH CONGENITAL HEART DISEASE
(EXCLUDING SITUS INVERSUS)

Cardiac abnormality	Other abnormality	Tribe	Sex
Atrial septal defect	Incomplete Marfan's disease	Ganda	M
Atrial septal defect	Mongol; bilobed rt. lung; common mesentery	Ganda	F
Atrial septal defect	Common mesentery	Ganda	F
Atrial septal defect and patent ductus arteriosus	Polydactyly; exomphalos	Ganda	F
Atrial septal defect and patent ductus arteriosus	Mongol; hydronephrosis	Ganda	M
Ventricular septal defect	Tracheo-oesophageal fistula	Ganda	F
Cor biloculare, patent ductus arteriosus supplies pulmonary artery, pulmonary vein joins superior vena cava	Cystic hygroma of tongue	Rwanda	M
Overriding aorta and pulmonary artery, atrial septal defect, patent ductus arteriosus, ectopic heart	Missing sternum; webbed neck; talipes equinovarus	Ganda	M
Tetralogy of Fallot	Mongol	Ganda	F
Aortic valve atresia, atrial and ventricular septal defects, common truncus	Spina bifida; fused legs; absent kidney	Gishu	M
Patent ductus arteriosus and coarctation	Polydactyly; syndactyly	Ganda	M
Persistent left superior vena cava	Anal atresia; hydronephrosis; cloacal hind gut; abnormal vertebrae; absent umbilical vein	Lugbara	?

TABLE III

TRIBAL DISTRIBUTION OF CASES OF CONGENITAL HEART DISEASE
(PERCENTAGES IN PARENTHESES)

Tribe	All ages	12 months of age and less		
	No. with congenital heart disease	No. with congenital heart disease	All necropsies	No. with congenital heart disease per 100 necropsies
Ganda	32 (54)	20 (53)	296 (41)	6.7
Rwanda	4 (6.7)	2 (5.3)	115 (16)	1.75
Luo	6 (10)	4 (10.6)	32 (4.4)	12.5
Ankole	4 (6.7)	3 (7.9)	28 (3.9)	10.7
Toro	3 (5)	2 (5.3)	13 (1.8)	15.4
Other	10 (17)	7 (18.5)	147 (20)	4.75
Not known	1 (1.7)	—	95 (13)	—
Total	60	38	726	5.25

In 7 of these the cardiac abnormality was, or included, an atrial septal defect. There was no other clear pattern of disorder.

Africans from Uganda, Kenya, and Tanzania contributed 58 of the cases of congenital heart disease—there was one Sudanese and one British case. Of the total of 60 cases, 32 (54%) belonged to the Ganda tribe, 6 to the Luo tribe, and 4 each to the Ankole and Rwandan tribes; the others came from 8 different tribes. When the 38 cases in children aged 12 months or less were considered alone, 20 (53%) were Ganda, 4 belonged to the Luo tribe, and 3 and 2, respectively, to the Ankole and Rwandan tribes (Table III). When these children's cases were compared with the number of necropsies in the same age-group, the 38 cases were found among 726 post-mortem records (5.25 per 100). Of these the 20 Ganda were found among 296 Ganda necropsies (6.7 per 100), the 4 Luo from 32 necropsies (12.5 per 100), the 3 Ankole from 28 necropsies (10.7 per 100), and the 2 Rwandan from 115 necropsies (1.75 per 100). These percentage differences, though large, do not reach conventional ($p < 0.05$) levels of significance.

Atrial septal defect was the dominant abnormality in 12 cases, occurring alone in 8 and in association with a patent ductus arteriosus in 4 infants less than 1 week old. Only 2 patients died of cardiac failure. The sex incidence was equal. The Ganda contributed 7 cases and no other tribe more than one. The frequency of associated abnormalities was high and has been mentioned. Atrial septal defect occurred with a single other significant abnormality in 2 adults with mitral stenosis (Lutembacher's syndrome), in 1 child with pulmonary valve stenosis, and in 2 children with anomalous pulmonary drainage, which was total in one and partial in the other. Atrial septal defect occurred with two or more other abnormalities which were of greater importance in a further 8 patients who are described under other headings.

Ventricular septal defect occurred alone in 8 patients, and with a patent ductus arteriosus in one other newborn case. Six of these patients were male, 3 belonged to the Luo tribe, and only 2 were considered to have died of cardiac failure.

Single ventricle was found 3 times. In one, there was also partial absence of the atrial septum

TABLE IV
CARDIAC ABNORMALITIES ASSOCIATED WITH VALVE ATRESIA

Atretic valve	Age	Sex	Tribes	Abnormalities
Mitral	7 dy.	M	Ganda	Wide patent ductus; high ventric. septal defect; large atrial septal defect; hypoplastic lt. atrium, lt. ventricle, and proximal aorta
Mitral	13 dy.	F	Ganda	Lt. ventricle only a diverticulum; rudimentary lt. atrium; large atrial septal defect; common truncus arteriosus; single coronary artery arises from rt. common carotid
Aortic	3 dy.	M	British	Patent ductus; wide atrial septal defect; lt. ventricular endocardial fibrosis
Aortic	1 dy.	M	Gishu	Atrial septal defect; low ventric. septal defect; common truncus arteriosus
Tricuspid	3 mth.	F	Ganda	Atrial septal defect; low ventric. septal defect; patent ductus; coarctation; transposition of aorta and pulmonary artery arising from lt. ventric., incomplete differentiation of pulmonary valve
Tricuspid	10 mth.	M	Luo	Single ventricle, large atrial septal defect

and a large patent ductus. The other two both had complete absence of the atrial as well as the ventricular septum. In addition, one had an abnormal pulmonary artery supplied through a large patent ductus arteriosus, while the other showed transposition of the aorta and pulmonary artery, patent ductus arteriosus, and an abnormal aortic valve.*

An overriding aorta was associated with ventricular septal defect in 2 patients, and there were 2 examples of the tetralogy of Fallot. Of these 4 patients, 3 were female Ganda children, one of whom was a mongol. A further Ganda female patient with overriding aorta and ventricular septal defect had in addition a large atrial septal defect, a large patent ductus, a heart exposed by the absence of the sternum, and webbing of the neck with talipes equinovarus.

Valve atresia was found in 6 children, with 2 cases each of tricuspid, mitral, and aortic atresia. Five of them had both atrial and ventricular septal defects and 2 had a common truncus (see Table IV). The sex incidence was equal, the Ganda contributed 3 patients, and other congenital abnormalities were present in only one case.

Patent ductus arteriosus alone was found 4 times and coarctation of the aorta alone once. Coarctation with patent ductus arteriosus was found in 3 children, and with a wide atrial septal defect as well in another. All the coarctations were preductal. Of the 4 with solitary patent ductus arteriosus, 3 were female, but if all 8 are considered, 4 were male and 4 were female. Patent ductus and coarctation were also found among multiple abnormalities in one of the patients with tricuspid valve atresia.

Valve cusp abnormalities were found in 5 children, with one example each of fenestrated aortic

cusps, fenestrated aortic and pulmonary cusps, absent pulmonary valve cusp, enlarged aortic valve cusp with an interventricular septal aneurysm and complicating bacterial endocarditis, and an angiomatous abnormality of the posterior cusp of the mitral valve. There was nothing in the first 3 cases to suggest bacterial endocarditis. All 5 were male, and death was due to pneumonia in 2 and pulmonary tuberculosis in 2.

Persistent left superior vena cava was found in a newborn child with gross non-cardiac abnormalities. The other cases of congenital heart disease (2 situs inversus, 2 fibro-elastosis, and one subaortic stenosis) all died of diseases unrelated to the cardiovascular system, and had no other congenital abnormalities.

DISCUSSION

The conclusions which can be drawn from a study of this kind are limited. Examples have been found of valve cusp abnormalities, situs inversus, fibro-elastosis, subaortic stenosis, and persistent left superior vena cava, which are conditions not previously reported in tropical Africa but which are well recognized elsewhere. No unique disorder or combination of abnormalities was recognized, and the only notable absentee from the list of conditions is transposition of the great vessels, which, however, has been recorded from tropical Africa by Caddell and Connor (1966) and also by Gupta and Antia (1967).

Details of the selection of cases for necropsy are unknown, so that no proper comparison can be made of the incidence of congenital heart disease in different tribes. However the relative frequency among the Luo infants and the relative infrequency among the Rwandans, when compared with the Ganda, hint at possible tribal differences, and further hints are that 3 of the 6 Luo patients had solitary ventricular septal defects, and that 4 of the 5 patients with overriding aorta were female Muganda children. Larger numbers of patients will have to be studied

* The post-mortem records of the second and third cases were incomplete, with inadequate descriptions of the valves. Inclusion under single ventricle is therefore somewhat arbitrary.

to confirm or refute these hints, but if tribal differences of incidence and pattern could be established, some light might be shed on the aetiology of congenital heart disease.

SUMMARY

Sixty cases of congenital heart disease are described, including 11 with conditions not previously reported from tropical Africa. No unique disorders were recognized. The tribal distribution has been recorded, and the results, though not

statistically significant, hint at tribal differences of incidence and pattern of congenital heart disease.

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