WHY THEY SURVIVE? NATIVE CONGENITAL HEART DISEASE IN A GROWN-UP AFRICAN POPULATION WITHOUT ACCESS TO CARE

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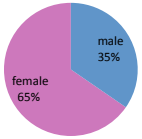
Background

In developed countries most children with congenital heart disease (CHD) are detected in the first months of life (or prenatally) and receive life saving heart surgery in the first year. Survival from CHD is now over 90% and in Europe, Japan and North America there are now more adults than children with CHD. Not so in less well resourced nations where access to diagnostic services is poor and management capacity weak . Before 2009 there were no cardiac services in Namibia thus we witness interesting insights into natural history of CHD. We describe a case series of 104 patients presenting with CHD over 13 years of age and with no previous treatment.



Gender

Methods

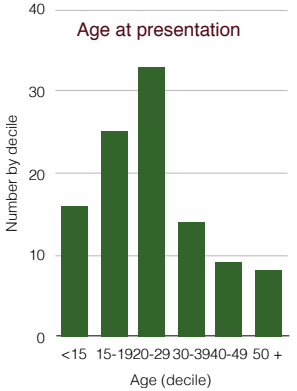
Case notes of all patients seen at Windhoek Central and Roman Catholic Hospital between January 2009 and December 2013 aged over 13 years were reviewed for congenital heart disease. Clinical features, ECG, chest x-ray, echocardiogram, cardiac catheterisation, operation reports and follow-up records were checked. Diagnosis was established by echocardiogram. Surgery and/or intervention was conducted at Windhoek Central Hospital, Roman Catholic Hospital in Namibia or, at Christiaan Barnard Memorial Hospital in South Africa.

Results

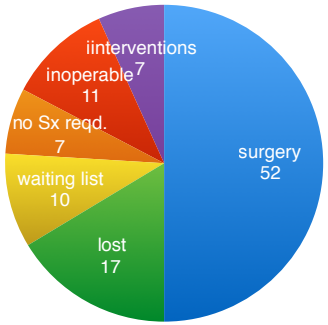
One hundred four (104) patients were identified with age range from 13 to 86 years (mean 25). 68 were female.

Primary diagnosis included, ventricular septal defect (VSD 30), secundum atrial septal defect (ASD) 32, patent arterial duct (PDA) 12, atrio-ventricular septal defect (AVSD) 10 (9 primum ASD), aortic stenosis 4 and coarctation of aorta 4. Three patients had Shone syndrome with coarctation, aortic and mitral stenosis.

Cyanotic disease included Tetralogy of Fallot 7 (TOF), double outlet right ventricle (DORV) with TOF 4, pulmonary stenosis (PS) 7, Ebstein's anomaly 2 and transposition of the great arteries with VSD/PS 2. Diagnostic cardiac catheterisation was performed in 27 the majority in whom to assess reversibility of pulmonary hypertension and severity of pulmonary vascular disease (14) or, for pulmonary anatomy in Tetralogy of Fallot (9). Eleven were inoperable (8 with Eisenmenger syndrome). Surgery or intervention was not necessary in only 7 patients, 5 of whom had restrictive VSD's.



Primary diagnosis	Number	Surgery	Intervention	Inoperable	Lost
Atrial septal defect	32	16	2	2	2
Atrioventricular septal defect	10	5		1	
Ebstein's anomaly	2	2			
Ventricular septal defect	30	15		3	5
Pulmonary stenosis	7	4	1		1
Tetralogy of Fallot	7	3			3
Double outlet right ventricle with TOF	4	3			
Double outlet right ventricle with VSD	1			1	
Patent arterial duct	12	5	2	1	3
Coarctation of the aorta	4	2	1		1
Aortic stenosis	3	3			
Sub-aortic stenosis	1	1			
Transposition great arteries/VSD/PS	2	1		1	
Tricuspid atresia	1			1	



There were 6 interventions for, coarctation of the aorta (1 pre-mounted covered CP stent), PDA (2 with Amplatzer ADO1 devices), ASD (2 with Amplatzer ASD closure devices) and valvular PS (1 Tyshack balloon valvuloplasty).
Seventy eight were "operable". Surgery was performed on 52, ten (10) were waiting for surgery and fifteen patients have been lost to follow-up.
Over 4 year follow-up (mean 32 months), three patients have died. Two with inoperable secundum ASD and one after surgery for TOF with pulmonary atresia.

Discussion:

- High numbers (74) with PDA, ASD and VSD is not surprising. The severity of their illness is but reflects the referral nature of the service.
- AVSD with only an ostium primum ASD (the bridging leaflets closing the "VSD"), is a predictor for survival. The oldest patient (86) had a primum ASD.
- Contrary to the literature, survival with TOF is not uncommon and they presented late with the mean age 20 years (range 13 to 29 years). Data from this small cohort suggests pulmonary artery index is a predictor for long term survival in TOF or, DORV with TOF. It is completely speculative whether this cohort with TOF had mild infundibular pulmonary stenosis and were therefore not cyanosed in early life.
- The two patients with Ebstein's anomaly both presented in the third decade with virtually complete atrialization of the right ventricle and both were suitable for replacement of the tricuspid valve. The numbers are two small to draw any conclusions regarding survival.
- Despite the fact they were counselled they needed intervention or surgery, a significant number (17) were lost to follow-up. This reflects a combination of factors including the novelty of cardiac services, large referral distances, poverty, low human development and weaknesses within the heath system.

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