Spectrum and age of presentation of significant congenital heart disease in KwaZulu Natal, South Africa

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KwaZulu Natal

- Population 10.3 million
- 3.3 million under 14 years
- 7th poorest province
- 9 districts
- 1 metropole -3.7 million people
- IMR: 32/1000

Paediatric Cardiac Services

- Public services : 85% of population
- 4 levels of care: District, Regional, Provincial Tertiary and Central
- 3 paed cardiologists based centrally
- Defined referral pattern
- 3 screening clinics at tertiary hospitals – run by paediatricians primarily

Challenges to services

- Late and Non referral of patients

- Capacity of service to handle patients that do present

Aims

- Snapshot of Paediatric Cardiac Services over one year period

- Patient characteristics
- Factors affecting late referral

- Outcomes

Method

- New patients seen in one calendar year –questionnaire on first presentation with informed consent
- Age at presentation
- Cardiac diagnosis
- Comorbidity/Risk factors
- Socioeconomic status
- Maternal Age and Education
- Access to Health Care
- Late presentation
  - Missed opportunities for diagnosis
  - Obvious symptoms/signs missed

Inclusion criteria

- **Significant congenital heart disease** presenting to IALCH.
  - Likely to require surgery in childhood
  - Included VSDs with initial large shunt

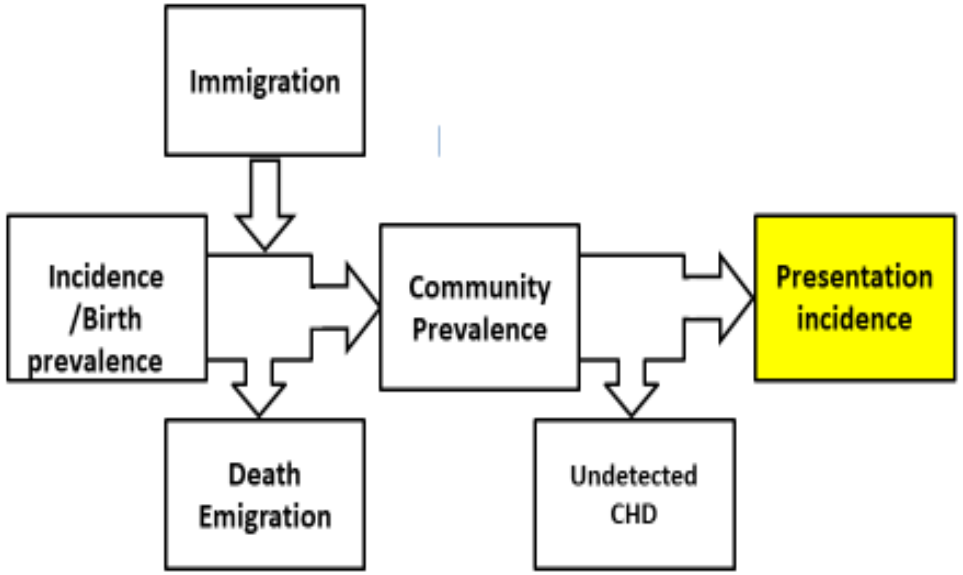
- Up to 12 completed years of life.

Exclusion criteria

- Previous palliative or correctional procedure
- Patent ductus arteriosus in a premature infant < 3 months of age
- Non-significant cardiac disease not meeting above definition

Defns and Terminology

- Variability between studies
- CHD incidence vs Birth prevalence vs prevalence
- Critical CHD – all infants dying or needing surgery within the first 28 days of life
- Severe CHD – needing surgery within the first year of life (CCHD in some studies)
- Significant congenital heart disease – requiring surgery in childhood



Classification of CHD

- Physiological rather than purely anatomical
- Most significant lesion if more than one lesion
- Multiple significant lesions counted as separate lesion

- Denominator: The number of live births recorded within the public service for the period under study : DHIS data file: 215,641 livebirths within the public sector

Results

- 316 pts
- 8 excluded – on review had not met inclusion criteria
- 14 not recruited – added with available information – predominantly ICU patients who arrived without caregivers

- Total 322 pts over one year

Parent socioeconomic characteristics

	Number	Total N	Percentage
Receiving/Eligible for Care Support Grant	263	322	81%
No clean tap water	38	313	12%
No electricity	37	313	11.8%
No water and electricity	19	313	6%
Number of mothers who completed secondary school	136	308	44%
Number who completed only primary school	30	308	9.7%
Post school diploma/degree	26	308	8.4%
Number of mums unemployed or scholars	248	322	77%
Both parents unemployed/scholars	129	322	40%
Number with paternal support or involvement	178	314	57%

HIV exposure

HIV status (n=322)		HIV Infected	HIV Uninfected	Infection status unknown	Maternal ARVs throughout pregnancy	Maternal ARVs after first trimester
HIV Exposed	113 (35.5%)	5 (1.55 %)	92	16	61 (19%)	42 (13%)
HIV Unexposed	206					
Unknown	3					

Comorbidity- Trisomy 21

- 58/322 patients with congenital heart disease had T21 (18% of total CHD)
- 43% of all T21 patients had dysmorphic features of T21 missed initially
- 51/58 (88%) presented after six months of age
- 50% had AVSD

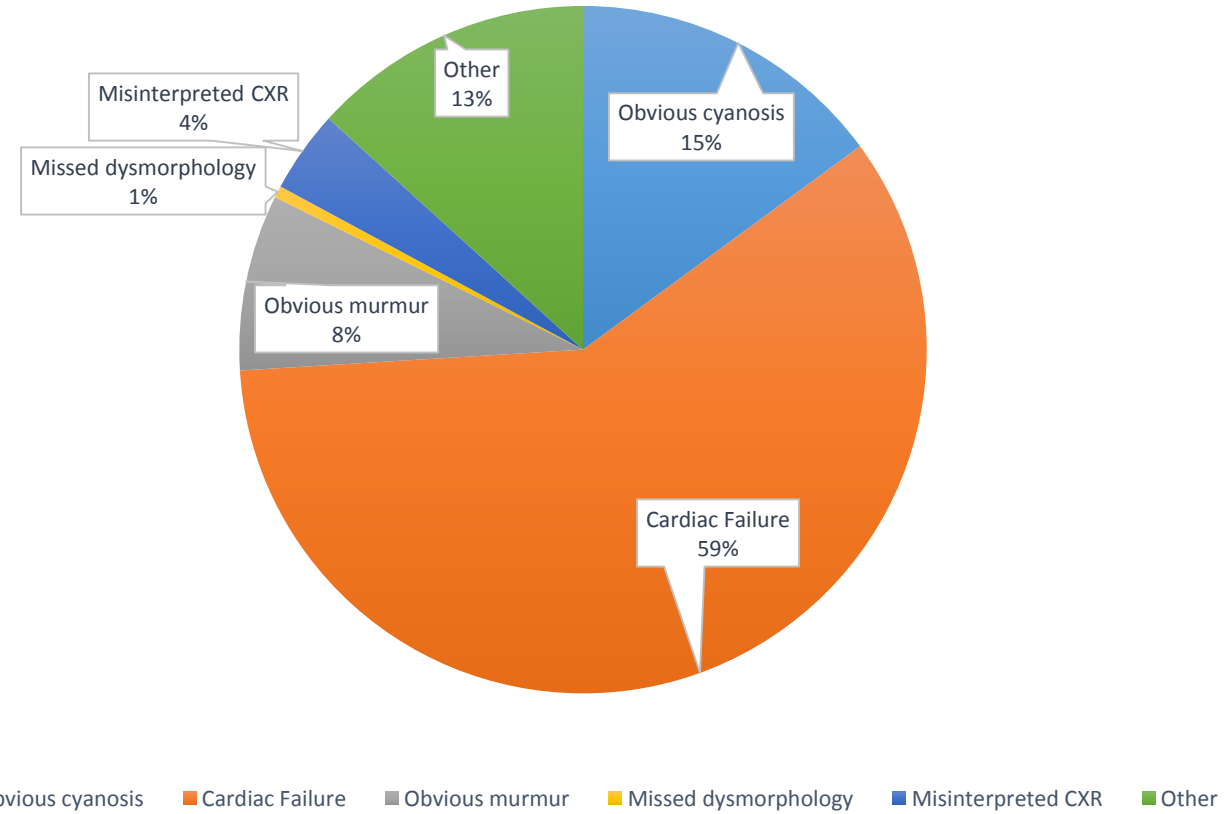
Most common CHD

	Total number of patients	Average age at presentation (yrs)	Range (days to yrs)	Median (Days/yrs)	Presentation after 6 months of age (%)	Presentation after one year of age(%)
ALL CHD	322	1.5	0 d to12.26 yrs	149/0.41	141(44%)	97(30%)
Isolated VSDs (PM and Muscular)	69 (21%)	1.22	2d to 8.1 yrs	126/0.35	28(41%)	17(25%)
Tetralogy (including PA/VSD)	57(18%)	2.39	2 d to 11.18yrs	204/0.56	30(53%)	25 (44%)
Patent Ductus Arteriosus	42 (13%)	1.06	8 days to 10.95 yrs	176/0.48	20 (48%)	15 (36%)
Complete AVSD	37 (11.5%)	0.69	6 days to 6.12 yrs	149/0.41	20 (54%)	15 (41%)
Secundum ASD	16 (5%)	3.19	66-3443/0.18-9.43	122/0.33	13(81%)	8 (50%)

	Total no. seen over 1 year	IALCH presentation incidence/1000	World birth prevalence meta-analysis Van der Linde et al	Incidence Hoffman et al (median)
All Significant CHD	322	1.61	6	7.67
VSD –all anatomical types	69	0.3		
ASD	16	0.074		
PDA	42	0.2		
Tetralogy	57	0.27	0.34	0.36
PS	14	0.07		
Coarctation	5	0.02	0.34	0.36
TGA	7	0.03	0.31	0.30
Aortic stenosis	4	0.01		
AVSD -complete	37	0.17	-	0.34

Late presentation

- “Mortality or morbidity which could have been avoided by earlier presentation and/or late presentation despite very obvious physical signs which should have brought the child to medical attention earlier”
- 55% - late presenters
  - 89% - obvious clinical signs missed
  - 35% - morbidity /mortality resulting from late presentation
  - 93% - missed at 6 week immunization visit
  - 43% - presented with symptoms related to cardiac condition – missed or misinterpreted initially – 39% seen by nurse only, 61% by a hospital doctor and 18% by a paediatrician
  - Increasing cost of travel to nearest health centre related to late presentation


Other
13%
Misinterpreted CXR
4%
Missed dysmorphology
1%
Obvious cyanosis
15%
Obvious murmur
8%
Cardiac Failure
59%
Obvious cyanosis
Cardiac Failure
Obvious murmur
Missed dysmorphology
Misinterpreted CXR
Other

Critical congenital heart disease

- “Any potentially life threatening duct-dependent disorder from which infants die or undergo invasive procedures (surgery or cardiac catheterisation) in the first 28 days of life “ as well as “all infants dying or needing surgery in the first 28 days of life with coarctation of the aorta, aortic valve stenosis, pulmonary valve stenosis, tetralogy of Fallot, pulmonary atresia with ventricular septal defect, or total anomalous pulmonary venous connection”

- Wren C, Reinhardt Z, Khawaja K Twenty-year trends in diagnosis of life-threatening neonatal cardiovascular malformations Archives of Disease in Childhood - Fetal and Neonatal Edition 2008;93:F33-F35.
- Thangaratinam S BK, Zamora J, Khan KS, Ewer AK. Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. Lancet. 2012;379:2459-64.

Cardiac Lesion	Number per category	Number seen	Prevalence/1000	Hoffman
All duct dependent systemic circulation	4		0.019	
Duct dependent coarctation/Interruption		3	0.014	
Hypoplastic Left Heart Syndrome		1	0.005	0.226
All duct dependent pulmonary circulation	11		.05	
Critical PS		1	0.005	
Tetralogy /PA/VSD		7	.032	
Tricuspid atresia/PS		2	0.009	
TGA /VSD/PS		1	0.005	
Cyanotic with increased PBF	3			
Simple TGA		2	0.009	0.30
TAPVC		1	0.005	0.91
TOTAL		18	0.13	

Limitations

- Assumes referral patterns are followed
- Live births as denominator
- Antenatal diagnosis and TOP

Conclusions

- Present health system in KZN is not geared for detection of congenital heart disease
- CHD becomes an increasingly important cause of child mortality as IMR/CMR improves
- Outcomes and regional differences in pick up rates additional part of study