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SCREENING FOR CONGENITAL HEART DISEASE

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INTRODUCTION

- incidence
 - prevalence varies
 - 1/100 to 8/1000
 - do not take into account e.g.
 - sub AS (10- 12/1000)
 - acquired HD (RHD – 15-25/1000)
- 85% should reach adult life (intervention)
- MRC statistics (www.mrc.ac.za/policybriefs/childmortality)
 - CHD responsible for 1,2% under 5 mortality
 - 8th most important cause of mortality in children
 - underestimated in SA!
 - probably more – Resp infection, GE

BURDEN OF PEDIATRIC HEART DISEASE: SOUTH AFRICA: PREVALENCE 2004

- SA
 - population: 42,7 mil
 - % children < 16: 6 834 965
 - RHD 7 518
 - CHD 8 885
 - both 16 403

HIV – exposed & infected ??

ONLY 40% children with CHD are receiving the care they should be getting

CHILDREN AT RISK OF HEART DISEASE

- intrauterine growth retardation
- low birth weight
 - SA: 12 -22%
- prematurity
- discharge < 2days of age
- chromosomal abnormalities
- multiple malformations
- specific lesions
 - CoA
 - Truncus
 - Ebstein
 - HLHS
 - ASD

NEONATAL CONGENITAL HEART DISEASE

- 12 per 1000 live births
 - + 5% muscular VSD's (ECHO)
- higher in developing world
 - high fertility rate > disproportionate number CHD
 - made worse by age structure of population
 - e.g. 857/million Nigeria vs 120/million Singapore
 - lower per capita income – worsens burden of disease/ wage earner
 - leads to more deaths CHD

- cost

TABLE 3. REPRESENTATIVE COSTS. DATA TAKEN FROM KUMAR AND BALAKRISHNAN⁴ AND PASQUALI *ET AL.*⁵

<i>Service</i>	<i>Africa</i>	<i>India</i>	<i>USA</i>
Open-heart surgery	US\$10 000	\$5 000	\$12 000–50 000
128-slice CT angiogram	-	\$350	\$4 000
Colour Doppler echocardiography	\$100	\$30	\$200

NEONATAL CRITICAL CONGENITAL HEART DISEASE

- definition
 - potentially life-threatening, systemic or pulmonary circulation ductal dependent
 - e.g. TGF,TA, PA, CoAo, HLHS
- undetected CCHD
 - 50% demise in hospital or home
 - 15% CHD (death , 12mo) – undiagnosed (UK)
 - present with CVS/RESP collapse after discharge

Why the concern?: TGA South Africa

TABLE 7: Annual paediatric cardiac surgical load for 2006

	Public sector hospitals					Private sector		
	Johannesburg	Durban	Bloemfontein	Pretoria	WCPCS	SH	CBMH	Unitas
CPB	90-100	100	133	76	140	249	~80-110	~45
Closed	20-30	30	22	65	90	68	~40-70	~30
RHD	15-20	38	10	15	25	16	<10	
Waiting list	>200	~150	26	nil	>200	10-20*		
Simple TGA operations	5	3	0	3	5	7	3	2

WCPCS: Western Cape Paediatric Cardiac Services; SH: Sunninghill Hospital; CBMH: Chris Barnard Memorial Hospital.
 CPB: cardiopulmonary bypass. Closed: operations without CPB. RHD: Rheumatic heart disease. TGA: Transposition of great arteries.
 *Applies to patients for Walter Sisulu Paediatric Cardiac Centre for Africa. ~ approximate annual numbers

TABLE 8: Summary of personnel, paediatric cardiology and paediatric cardiac surgical procedures performed annually in South Africa

	Public sector hospitals	Private sector
Paediatric cardiologists	12	12
Paediatric cardiac surgeons	8	5
Cardiology trainees	8	-
Echocardiograms	15 385+	5 637+
Total cardiac catheterisations	1 100	289+
Interventional cardiac catheterisations	353	~133
Operations for congenital heart disease	800	~570
Simple transposition of great arteries operated on in 2006	18	12
Operations for rheumatic heart disease	100	~20

**vs 114 estimated
= ≤ 30%!!!**

Summary for personnel is for end 2008. Other data is annual data for 2006 except where estimates were provided (indicated by ~). + indicates where procedures performed in the country as a whole are in excess of the number shown as procedures performed outside the major centres are not included or data from some centres were not supplied.

SCREENING MODALITIES

- clinical
- echocardiography
- pulse oximetry

CLINICAL SCREENING

- varied reports
- examination at birth – 50% detection
 - UK study – adequate training 70 – 80%
- small team examining pre-discharge
 - does not matter whether physician or registered nurse
 - experienced team
 - **structured referral**
 - continuing education

ECHOCARDIOGRAPHIC SCREENING

- echocardiography
 - improves detection
 - not routinely available
 - need expertise
 - expensive – reduce cost of OPD referrals
- J Perinatal Med 2002;30;307-12
- antenatal cardiac ultrasound
 - 20weeks
 - detection rate
 - average: 23%
 - range: 15 – 68%
 - advantages
 - early detection
 - delivery in high risk unit
 - termination
 - disadvantages
 - cost
 - skill
 - SA ?? (developing countries < 15%)

OXIMETRY

PULSE OXIMETRY

- how effective?

	Limb	Antenatal diagnosis of CHD	Test timing	Total	True positive	False positive	False negative	True negative	Sensitivity (%; 95% CI)	Specificity (%; 95% CI)	Likelihood ratio positive (%; 95% CI)	Likelihood ratio negative (%; 95% CI)	False-positive rate (%; 95% CI)
Meberg et al (2008) ³³	Foot only	Excluded	<24 h	50 008	27	297	8	49 676	77.1% (59.9-89.6)	99.4% (99.3-99.5)	129.8% (104.9-160.6)	0.23% (0.13-0.43)	0.6% (0.5-0.7)
Bakr et al (2005) ³⁵	Foot and right hand	Excluded	>24 h†	5211	3	2	0	5206	100.0% (29.2-100.0)	100% (99.9-100.0)	1823.1% (500.1-6646.1)	0.13% (0.01-1.67)	0% (0-0.1)
Arlettaz et al (2006) ³⁴	Foot only	Included	<24 h	3262	12	12	0	3238	100.0% (73.5-100.0)	99.6% (99.4-99.8)	250.1% (142.3-439.5)	0.04% (0.01-0.59)	0.4% (0.2-0.6)
Sendelbach et al (2008) ³⁶	Foot only	Excluded	<24 h	15 233	1	24	0	15 208	100.0% (2.5-100.0)	99.8% (99.8-99.9)	466.3% (191.0-1138.5)	0.25% (0.02-2.8)	0.2% (0.1-0.2)
Reich et al (2003) ^{37*}	Foot and right hand	Excluded	>24 h†	2114	0	4	0	2110	--	99.8% (99.5-99.9)	--	--	0.2% (0.1-0.5)
Koppel et al (2003) ³⁸	Foot only	Excluded	>24 h	11 281	3	1	2	11 275	60.0% (14.7-94.7)	100.0% (100.0-100.0)	6765.6% (839.8-54 506.3)	0.40% (0.14-1.17)	0% (0.0-0.0)
Rosati et al (2005) ³⁴	Foot only	Excluded	>24 h	5292	2	1	1	5288	66.7% (9.4-99.2)	100.0% (99.9-100.0)	3526.0% (424.6-29 282.9)	0.33% (0.07-1.70)	0% (0.0-0.1)
Richmond et al (2002) ³¹	Foot only	Included	<24 h	5626	8	56	1	5561	88.9% (51.8-99.7)	99.0% (98.7-99.2)	89.2% (62.9-126.3)	0.11% (0.02-0.71)	1% (0.8-1.3)
de Wahl Granelli (2009) ³⁹	Foot and right hand	Excluded	>24 h†	39 821	19	68	10	39 724	65.5% (45.7-82.1)	99.8% (99.8-99.9)	383.4% (268.8-546.9)	0.35% (0.21-0.57)	0.2% (0.1-0.2)
Riede (2010) ⁴¹	Foot only	Excluded	≥24 h	41 442	14	40	4	41 384	77.8% (52.4-93.6)	99.9% (99.9-99.9)	805.5% (542.0-1197.0)	0.22% (0.09-0.53)	0.1% (0.1-0.1)
Ewer et al (2011) ⁴²	Foot and right hand	Included	<24 h	20 055	18	177	6	19 854	75.0% (53.3-90.2)	99.1% (99.0-99.2)	84.9% (64.6-111.6)	0.25% (0.13-0.50)	0.9% (0.8-1.0)
Kawalec et al (2006) ³²	Foot only	Excluded	≥24 h	27 200	7	13	1	27 179	87.5% (47.3-99.7)	100.0% (99.9-100.0)	1830.2% (1001.2-3345.9)	0.13% (0.02-0.78)	0% (0.0-0.1)
Hoke et al (2003) ^{37*}	Foot and right hand	Included	<24 h	2876	4	53	0	2819	100.0% (33.8-100.0)	98.2% (97.6-98.6)	48.3% (30.6-74.7)	0.10% (0.01-2.48)	1.8% (1.4-2.4)
Summary estimate	--	--	--	229 421	--	--	--	--	76.5% (67.7-83.5)	99.9% (99.7-99.9)	549.2% (232.8-1195.6)	0.24% (0.17-0.33)	0.14% (0.06-0.33)

CHD=congenital heart defect. *Studies by Hoke and colleagues and Reich and colleagues excluded from the analysis. †Mean age at testing >24 h after birth.

Table: Accuracy estimates of primary studies for pulse oximetry in the detection of critical congenital heart defects in newborn babies

STUDY FINDINGS

- efficacy
 - ability to detect (sensitivity): 75%
 - accuracy (specificity): 99.2%
 - detected additional 30 cases per 100 000
- cost effectiveness
 - probability > 90%
- disadvantages
 - false positive rate
 - 0.05 (99,95 specificity)
 - parental anxiety
 - widely acceptable to parents

PRACTICAL ASPECTS

- how?
 - free from motion artefacts
- position of probe
 - hand
 - foot
 - hand + foot
- what saturation?
 - 95%
 - positive if:
 - $SO_2 < 90\%$ any time
 - $< 95\%$ both extremities
 - $> 3\%$ difference hand/foot x 3, each 1h apart
 - high altitude??
- when?
 - 24 -72 h $>$ least false positives

OXIMETRY

- lack of data in areas with high IMR
- feasible, easy test
 - no additional staff required
 - average time for screen = 3.5min
 - must understand some forms CHD **not** detected
- pulse oximetry will detect more CCHD
 - also other diseases – Resp
- requirements
 - integrate with current birthing services
 - infrastructure
 - training
- *comprehensive care should be available for infants*

Congenital Heart Disease (CHD) Assessment

09/11

Child in well-baby nursery 24-48 hours of age or shortly before discharge if < 24 hours of age

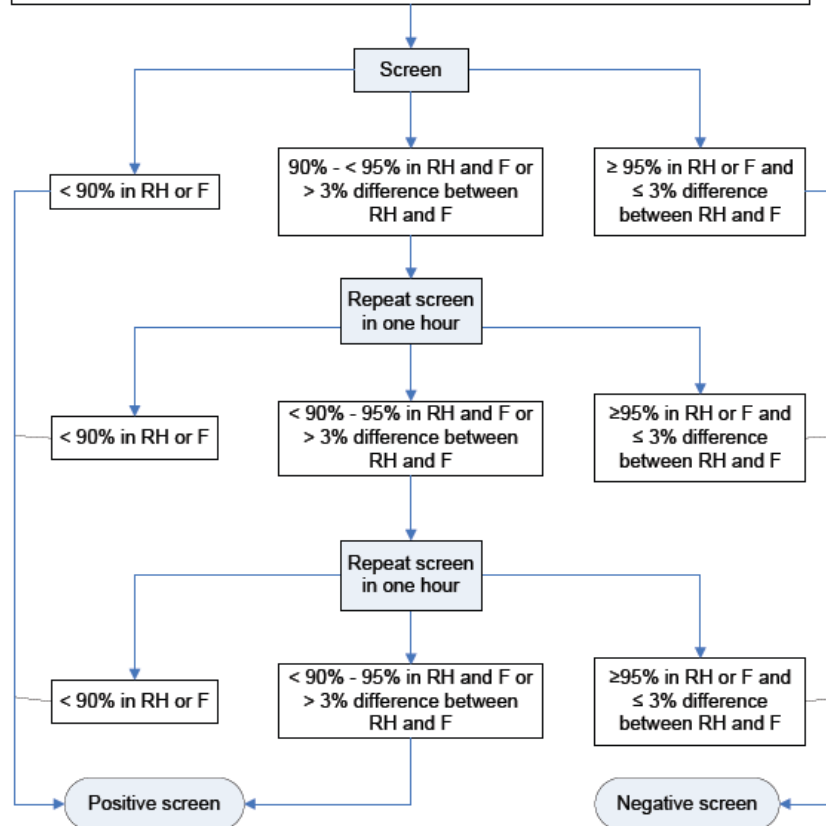


Figure. The proposed pulse oximetry monitoring protocol based on results from the right hand (RH) and either foot (F).

POTENTIAL SOLUTIONS

- MDG
- poverty = greatest barrier to successful Rx
- because Rx expensive
 - more attention to prevention
- prevention
 - education
 - family planning
 - involve local population
- **NB once IMR improves CHD becomes even MORE important**



“we have an obligation as uniquely talented individuals to change the boundaries of our thinking, the boundaries of our influence and the boundaries of our efforts”

Thank You
Dankie
Ke a leboha



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EPIDEMIOLOGY: CHD

- **incidence of CHD 4- 12/1000 live births**
- **85% should reach adult life**
- **CHD accounts for 50% of all deaths from *malformations***
- **50% severe – intervention**
- **MRC statistics** (www.mrc.ac.za/policybriefs/childmortality)
 - CHD responsible for 1,2% under 5 mortality
 - 8th most important cause of mortality in children
 - underestimated in SA !
 - probably more – Resp infection, GE

immediate outcomes

all survived

no major complication

1 balloon angioplasty

length of stay

patients 1- 4: 7, 2, 3 & 4 days

“we have an obligation as uniquely talented individuals to change the boundaries of our thinking, the boundaries of our influence and the boundaries of our efforts”

The Millennium Development Goals and Cardiovascular Disease

Cardiovascular disease is the leading cause of death worldwide despite the fact that the majority of cardiovascular disease deaths are preventable or treatable - The time has come to act!



GOALS AND TARGETS from the Millennium Declaration

1. ERADICATE EXTREME POVERTY AND HUNGER

Target 1B: Reduce extreme poverty and hunger. Halve the proportion of people living on less than \$1 per day by 2010.

Target 1C: Reduce the proportion of people who suffer from hunger.

CARDIOVASCULAR DISEASE AND THE MDGs

Cardiovascular disease (CVD) epidemic: There is now a worldwide epidemic of CVD, with the highest burden in men and middle-income countries. Every 30 seconds, someone dies from a CVD. In 2008, CVD was the leading cause of death worldwide. It is projected that by 2030, CVD will be the leading cause of death in all countries, including low-income countries.

SDG status: The world is not on track to meet the MDG targets for poverty reduction and hunger reduction. The proportion of people living on less than \$1 per day has not halved, and the proportion of people who suffer from hunger has not been reduced.

3. PROMOTE GENDER EQUALITY AND EMPOWER WOMEN

Target 3B: Remove all barriers to women's full participation in economic and social life.

CVD and education for women: The burden of CVD is disproportionately high among women in low-income and middle-income countries. Women with low education levels are at higher risk of CVD. Improving women's education and employment opportunities can reduce their risk of CVD.

4. REDUCE CHILD MORTALITY

Target 4: Reduce the under-five mortality rate to less than 7 per 1,000 live births by 2015.

CVD in the context of other maternal and child health: CVD is a leading cause of death in children and young adults. Improving maternal and child health can reduce the risk of CVD. For example, reducing maternal and child mortality can reduce the burden of CVD.

5. IMPROVE MATERNAL HEALTH

Target 5B: Reduce the global maternal mortality ratio to fewer than 100 per 100,000 live births by 2015.

Global trends in healthy pregnancies and safe births: The global maternal mortality ratio has declined by 45% since 2000. However, progress is uneven, and the goal of 100 per 100,000 live births is not yet met. Improving maternal health can reduce the risk of CVD.

8. DEVELOP ECONOMIC RESILIENCE

Target 8B: Double the world's gross fixed capital formation (GFCF) in infrastructure by 2015.

Healthcare, a high job sector for CVD, greatly increases the rates of completion during pregnancy for women: Women with low education levels are at higher risk of CVD. Improving women's education and employment opportunities can reduce their risk of CVD.

Advanced health insurance can be important for health care quality, thereby saving families in extreme financial hardship: Health insurance can protect families from financial hardship, which can reduce the risk of CVD.

Increasing safety net for vulnerable or informal workers: Informal workers are at higher risk of CVD. Improving their safety net can reduce their risk of CVD.

