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

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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

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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

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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 BALAKRISHNAN[4] AND PASQUALI *ET AL.*[5]

Service	*Africa*	*India*	*USA*
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

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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

Zuhlke SAHeart 2013
Chang Arch Ped Adol Med 2008
Abu-Harb Arch Dis Child 1994

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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

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.

vs 114 estimated
= ≤ 30%!!!

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SCREENING MODALITIES

- clinical
- echocardiography
- pulse oximetry

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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

Arch Dis Child Fetal Neonatal 2006;91:F263-7

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ECHOCARDIOGRAPHIC SCREENING

- echocardiography
  - improves detection
  - not routinely available
  - need expertise
  - expensive – reduce cost of OPD referrals

    J Perintal 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%)

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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)[30]	Foot only	Excluded	<24 h	50008	27	297	8	49676	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 ht	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	15233	1	24	0	15208	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)[31]*	Foot and right hand	Excluded	>24 ht	2114	0	4	0	2110	..	99·8% (99·5-99·9)	..	..	0·2% (0·1-0·5)
Koppel et al (2003)[29]	Foot only	Excluded	>24 h	11281	3	1	2	11275	60·0% (14·7-94·7)	100·0% (100·0-100·0)	6765·6% (839·8-54506·3)	0·40% (0·14-1·17)	0% (0·0-0·0)
Rosati et al (2005)[24]	Foot only	Excluded	>24 h	5292	2	1	1	5288	66·7 % (9·4-99·2)	100·0% (99·9-100·0)	3526·0% (424·6-29282·9)	0·33% (0·07-1·70)	0% (0·0-0·1)
Richmond et al (2002)[33]	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)[28]	Foot and right hand	Excluded	>24 ht	39821	19	68	10	39724	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)[37]	Foot only	Excluded	≥24 h	41442	14	40	4	41384	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)[27]	Foot and right hand	Included	<24 h	20055	18	177	6	19854	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)[38]	Foot only	Excluded	≥24 h	27200	7	13	1	27179	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)[32]*	Foot and right hand	Included	<24 h	2876	4	53	0	2819	100·0% (39·8-100·0)	98·2% (97·6-98·6)	48·3% (32·6-71·7)	0·10% (0·01-1·40)	1·8% (1·4-2·4)
Summary estimate	..	..	..	229421	..	..	..	..	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

Thangaratinam. Lancet 2012

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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

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PRACTICAL ASPECTS

- how?
  - free from motion artefacts
- position of probe
  - hand
  - foot
  - hand + foot
- what saturation?
  - 95%
  - positive if:
    - SO2 < 90% any time
    - < 95% both extremities
    - > 3% difference hand/foot x 3, each 1h apart
  - high altitude??
- when?
  - 24 -72 h > least false positives

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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*

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Congenital Heart Disease (CHD) Assessment

09/11

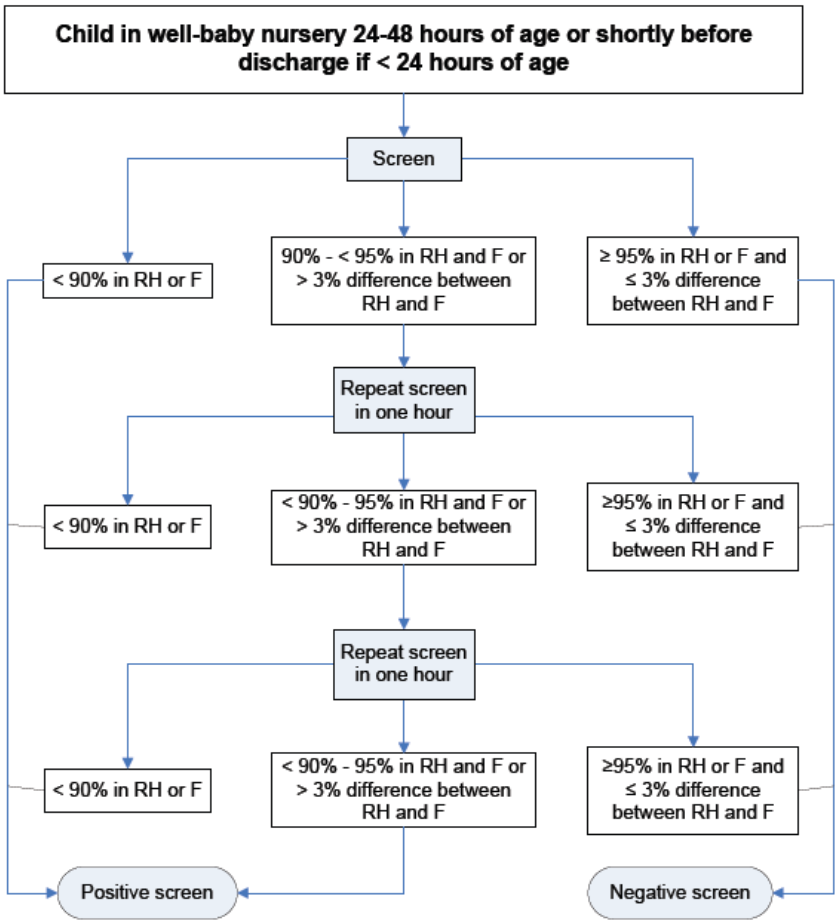

Figure. The proposed pulse oximetry monitoring protocol based on results from the right hand (RH) and either foot (F).

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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***

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“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

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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”

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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 to act is now!

GOALS AND TARGETS from the Millennium Declaration

CARDIOVASCULAR DISEASE AND THE MDGs