CHARACTERISTICS OF CHILDREN PRESENTING WITH CARDIOMYOPATHY IN AN AFRICAN SETTING – INITIAL FINDINGS OF THE IMHOTEP REGISTRY

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BACKGROUND

Cardiomyopathy remains a leading cause of morbidity and mortality in children worldwide. Although we see 30 – 40 incident cases annually in Cape Town, the characteristics of childhood cardiomyopathies in African children have not been systematically described.

IMHOTEP is the first prospective, open-ended registry of prevalent and incident cases of heart muscle disease in children and adults in Africa. It will expand from pilot sites in Cape Town to other centres in South Africa and subsequently the whole continent to accurately characterize cardiomyopathies in Africa. This report describes the experience to date from the first paediatric sites.

METHODS

Commencing August 2016, all incident cases of cardiomyopathy or myocarditis presenting to two referral paediatric cardiac centres were enrolled into a dedicated OpenClinica registry (IMHOTEP African Cardiomyopathy and Myocarditis Registry).

Details captured include demographics, history, physical examination, blood investigations, genetic screening, chest X-ray, ECG, echocardiogram features and outcome measures (adverse events, hospitalizations, death).

Children were enrolled at Red Cross Children's and Tygerberg Hospitals.

RESULTS

Baseline demographic characteristics, modes of initial presentation, and adverse outcomes are depicted in the tables.

In total, 11/25 (44%) of the patients are infants. Four of the 25 patients (16%) are perinatally HIV exposed, but all four tested negative by PCR.
The predominant cardiomyopathy phenotype is dilated cardiomyopathy − 12/25 (48%) followed by myocarditis (biopsy-proven, suspected clinically or on CMR) − 7/25 (28%).
Morbidity and mortality to date has been substantial – seven deaths (mortality 28%), average hospitalizations 2.7/patient, average ICU admissions 1.3/patient, and mean number of days in hospital 28.6 (SD 17).

Patient Demographics

Patient	Age (m)	Sex	Caregiver education	HIV status
1	10	F	High school	NE
2	1.5	M	High school	NE
3	6	F	Primary school	E-neg
4	113	M	Postgraduate	NE
5	2.5	M	Unknown	E-neg
6	32	M	High school	NE
7	3	M	Tertiary	NE
8	108	F	Tertiary	NE
9	5	F	High school	NE
10	3	F	Tertiary	NE
11	89	M	High school	Unknown
12	7.5	M	Primary school	NE
13	0.6	F	Tertiary	NE
14	0.6	F	Tertiary	E-neg
15	0.3	M	High school	E-neg
16	47	F	Tertiary	NE
17	156	F	High school	NE
18	16	M	High school	NE
19	30	M	Unknown	NE
20	35	F	High school	NE
21	13	M	High school	NE
22	24	F	Unknown	NE
23	99	F	Unknown	NE
24	79	F	Unknown	NE
25	149	F	High school	NE

NE – non-exposed, E-neg - exposed, PCR negative

Initial Presentation

Patient	Diagnostic category	Modified Ross class	Highest level of care	Days in ICU	Days in hospital at first admission
1	Myocarditis	IV	ICU	4	10
2	DCM	III	High care	0	4
3	DCM	IV	ICU	3	7
4	Myocarditis	II	ICU	9	12
5	DCM (due to EAT)	III	ICU	8	24
6	DCM	III	Ward	0	25
7	HCM	II	Ward	0	1
8	DCM	III	ICU	9	27
9	DCM	IV	ICU	3	10
10	DCM	II	Ward	0	8
11	RCM	II	Ward	0	8
12	DCM	III	ICU	11	18
13	LVNC	IV	ICU	7	14
14	LVNC	IV	ICU	9	14
15	Myocarditis	IV	ICU	4	11
16	Myocarditis	I	Ward	0	16
17	DCM	IV	ICU	28	28
18	Myocarditis	IV	ICU	7	15
19	DCM	IV	ICU	2	17
20	DCM	II	Ward	0	4
21	DCM	IV	ICU	23	24
22	Myocarditis	IV	ICU	14	46
23	DCM (due to EAT)	IV	ICU	12	26
24	DCM	III	Ward	0	2
25	Myocarditis	IV	ICU	3	9

DCM, HCM, RCM – dilated, hypertrophic, restrictive cardiomyopathy respectively,
LVNC – left ventricular non-compaction,
EAT – ectopic atrial tachycardia

Adverse Events / Outcomes*

Patient	Hospitalizations (total no.)	ICU admissions	Total days in hospital	Alive /died
1	7	3	62	Alive
2	4	3	53	Alive
3	3	2	20	Died in ward
4	1	1	12	Alive
5	2	1	24	Alive
6	1	0	25	Alive
7	3	0	3	Alive
8	3	1	52	Alive
9	3	3	62	Died in ICU
10	5	1	28	Died in ICU
11	4	0	24	Alive
12	2	2	23	Alive
13	4	2	30	Died in ICU
14	3	2	37	Died in ICU
15	1	1	11	Alive
16	2	0	17	Alive
17	1	1	28	Died in ICU
18	2	1	15	Alive
19	2	2	21	Alive
20	1	0	4	Alive
21	5	3	55	Died in ward
22	4	1	55	Alive
23	1	1	26	Alive
24	2	0	20	Alive
25	1	1	9	Alive

*Current to July 2017

CONCLUSIONS

- Already in these early stages of the registry, we see the full range of phenotypes represented in African children with cardiomyopathy and a significant mortality and morbidity.
- The paediatric limb of the IMHOTEP African Cardiomyopathy Registry promises to be a powerful tool to characterize childhood cardiomyopathy in Africa.

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