

Cardiomyopathy in children in the Western Cape: initial findings of the IMHOTEP registry

George AM Comitis
Division of Paediatric Cardiology
Department of Paediatrics & Child Health
University of Cape Town
SA Heart, 6 October 2018



Background

- Annual incidence 1.13 per 100 000 children, infants 8.34 per 100 000 (Ped CMO Registry - US)
- RXH ICU study – 95 children with fulminant CMO or myocarditis 2010-2015, 53% mortality (B. Rossouw)
- Outcomes: traditional teaching – 1/3, 1/3, 1/3 - ?true
- PCMR data (n = 741) – idiopathic DCM at 2 years:
 - 22% recovered normal LV function and size;
 - 27% had persistently abnormal echocardiograms;
 - 51% died or transplanted (median 3.2 months)
 - Predictors of recovery: younger age, less LV dilation

Pediatric Cardiomyopathy 2018 Fast Facts



A classroom of
25
children are
diagnosed
with cardiomyopathy
every week
in the United States

**LEADING
CAUSE**
of **SUDDEN
CARDIAC
ARREST**
in the young

Mortality rate
EXCEEDS
mortality of
all childhood
**CANCERS
COMBINED**



2 out of 3 children
do not have a known cause

Out-of-pocket treatment
expenses can add up to **\$62,000** per year

47 research studies on childhood cancer
to **1** study on pediatric cardiomyopathy

For every \$100
of government
spending on heart
disease research

65¢
goes to pediatric
cardiomyopathy
research



40%
of all diagnosed children
will either receive a
heart transplant or die

A Cause for Today... A Cure for Tomorrow
866.808.CURE • childrenscardiomyopathy.org



Epidemiology in Africa

- No paediatric data
- Adults
 - Viruses (including HIV), peripartum, other infections
 - Rate of myocarditis as a percent of heart failure: 0–50%
 - DCM – major cause of heart failure but myocarditis rarely identified (lack of available accurate diagnostic tools)

Role of cardiovascular MRI

- Most helpful noninvasive tool for diagnosis
- Best if done within 2 weeks of symptom onset
- Combination of T2-weighted and post-gadolinium early and late T1-weighted → best sensitivity (67%), specificity (91%) and accuracy (78%)

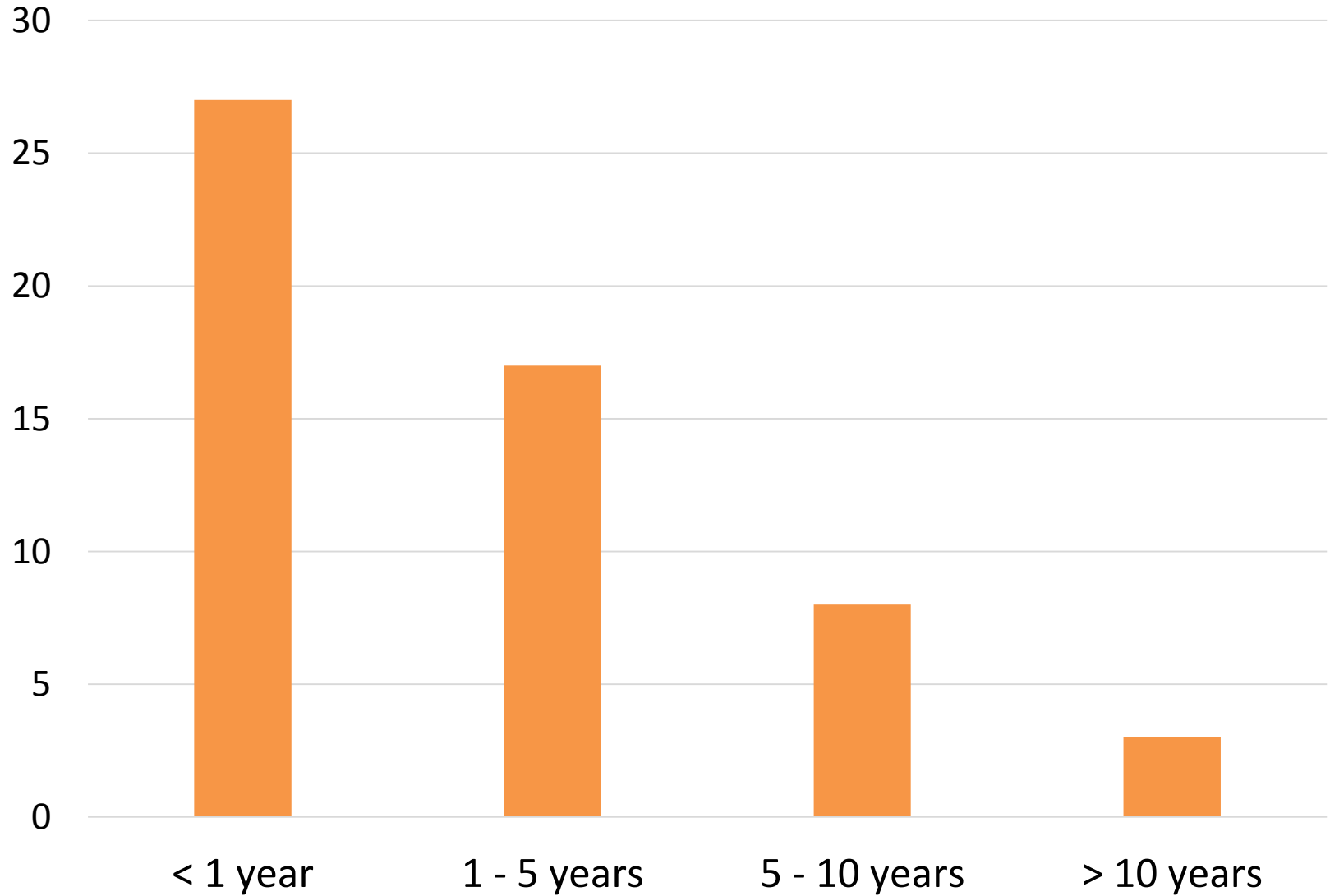
Methods

- The African Cardiomyopathy and Myocarditis Registry Program (IMHOTEK) – prospective, open-ended registry, pan-African, multi-center, hospital-based cohort study
- Pilot sites in CT → SA → Africa-wide
- Aim to systematically characterise CMO in African children:
 - Demographics, history, physical examination
 - Blood investigations, CXR, ECG, echo, genetic screening, CMR
 - Outcomes (adverse events, hospitalizations, death)
- From Sept 2016, all incident cases at RXH/TBH enrolled into OpenClinica database registry

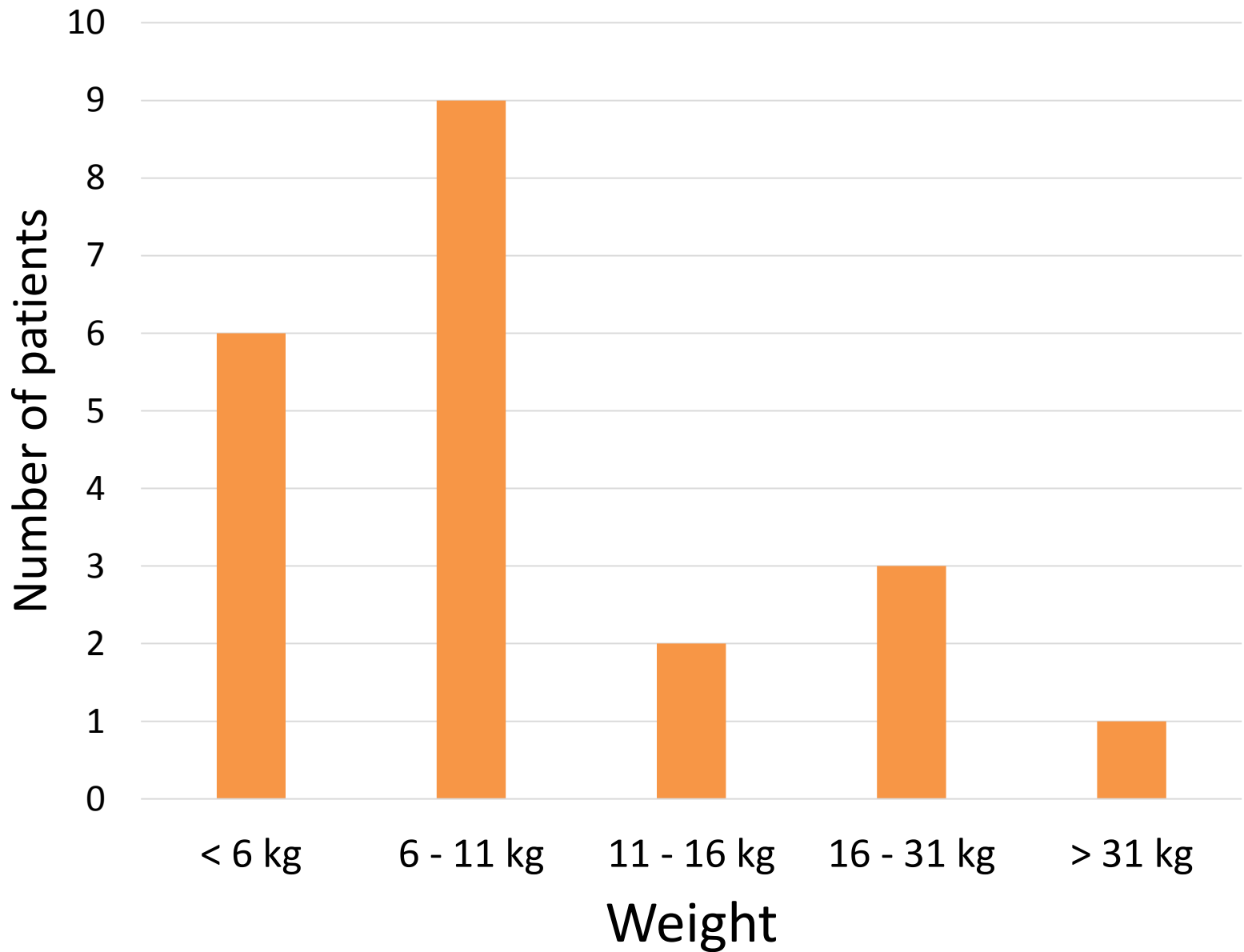
Results

- 56 children recruited up to September 2018
- 27 (48%) infants
- Mortality 15 (27%)
- CMO phenotype
 - dilated 31 (55%)
 - myocarditis 14 (25%)
 - LV non-compaction 4 (7%)
 - hypertrophic 4 (7%)
 - tachycardia-induced CMO 2 (3.5%)
 - restrictive 1 (2%)

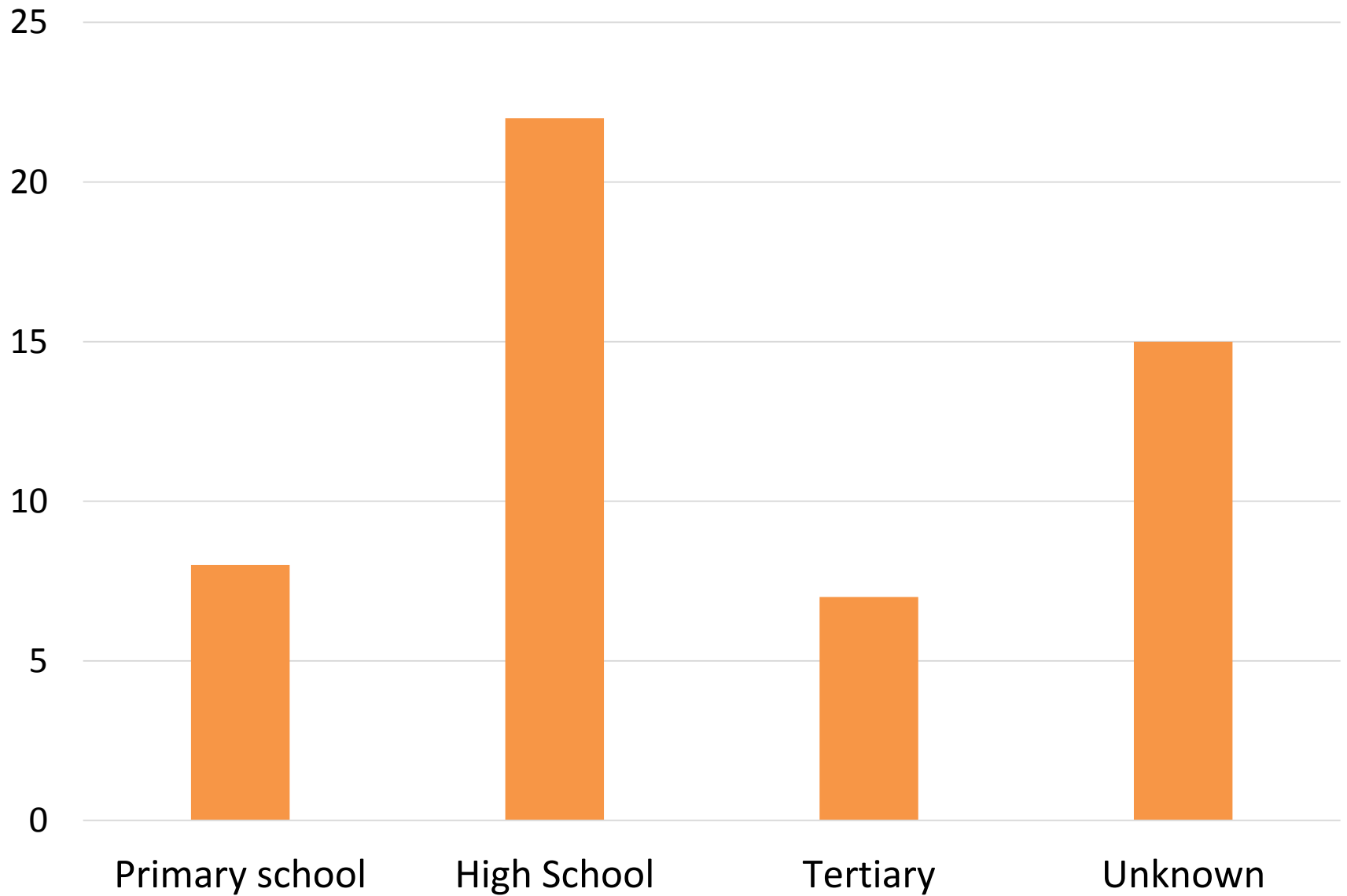
Age at first presentation



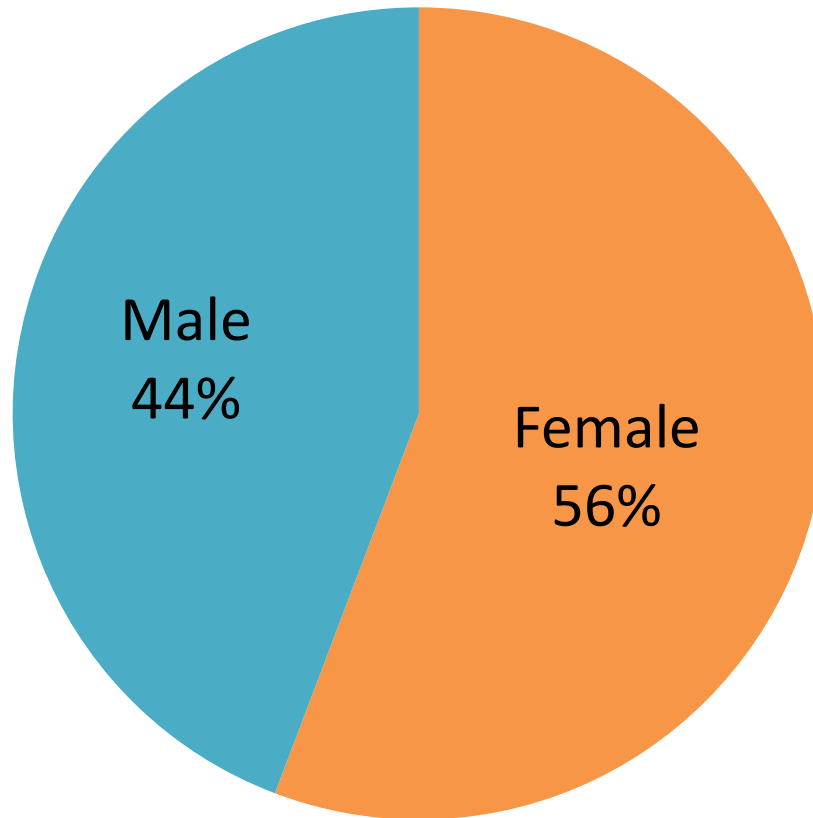
Weight distribution



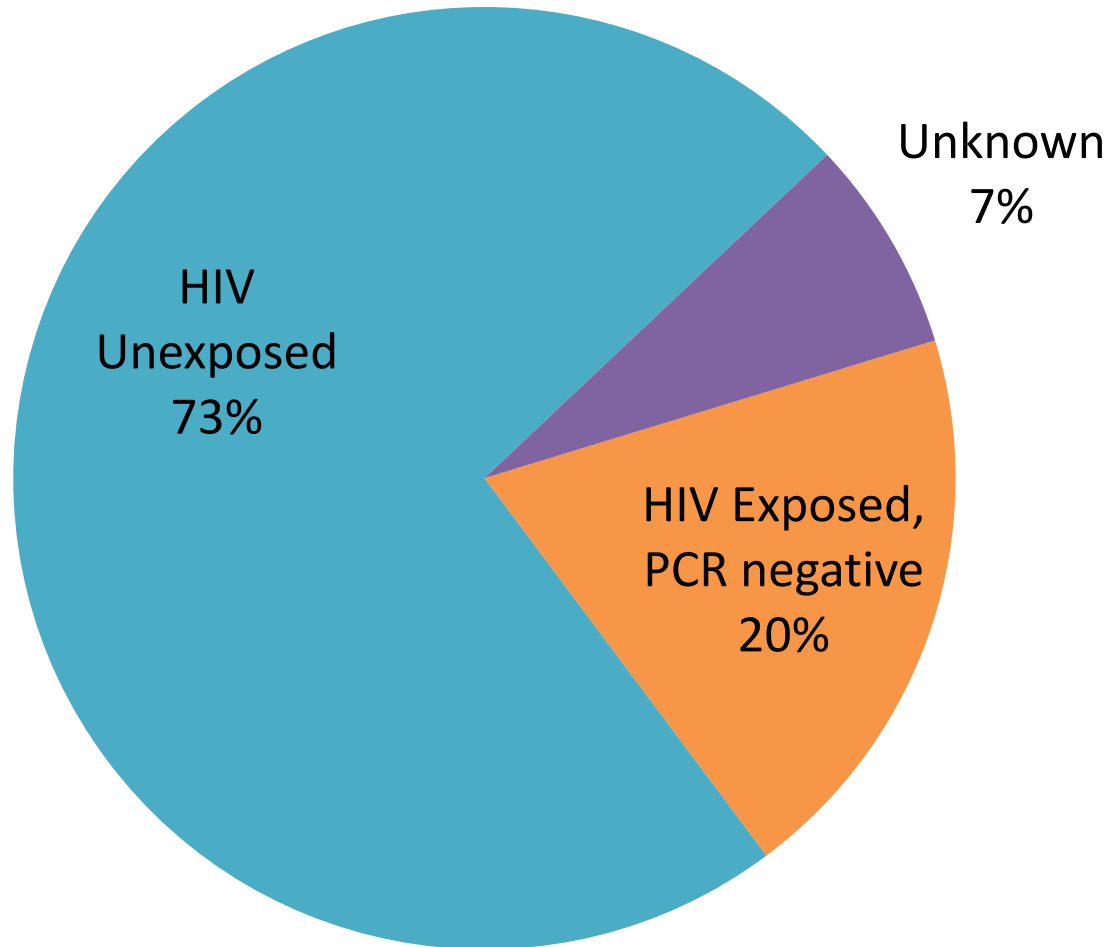
Maternal education



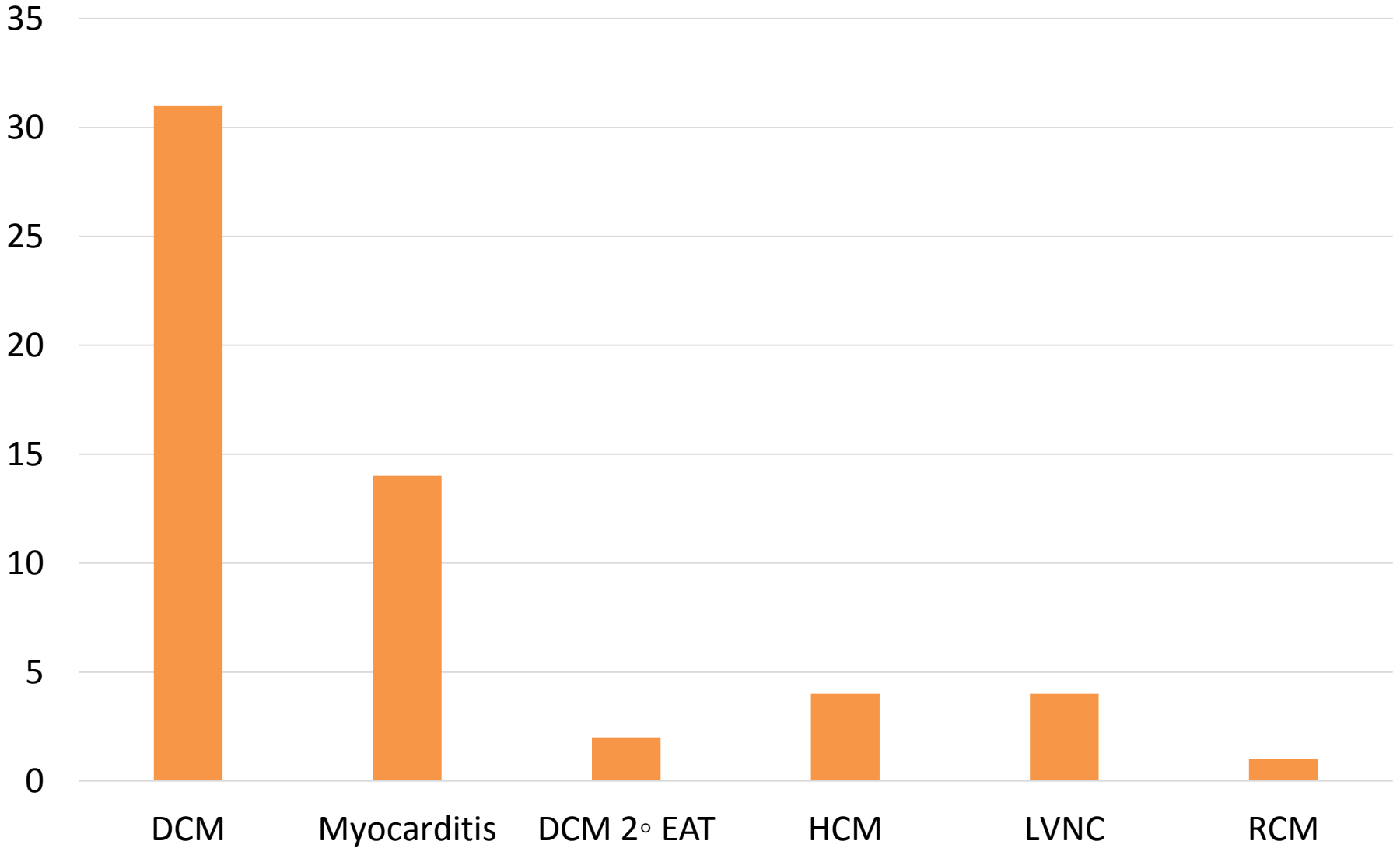
Gender distribution



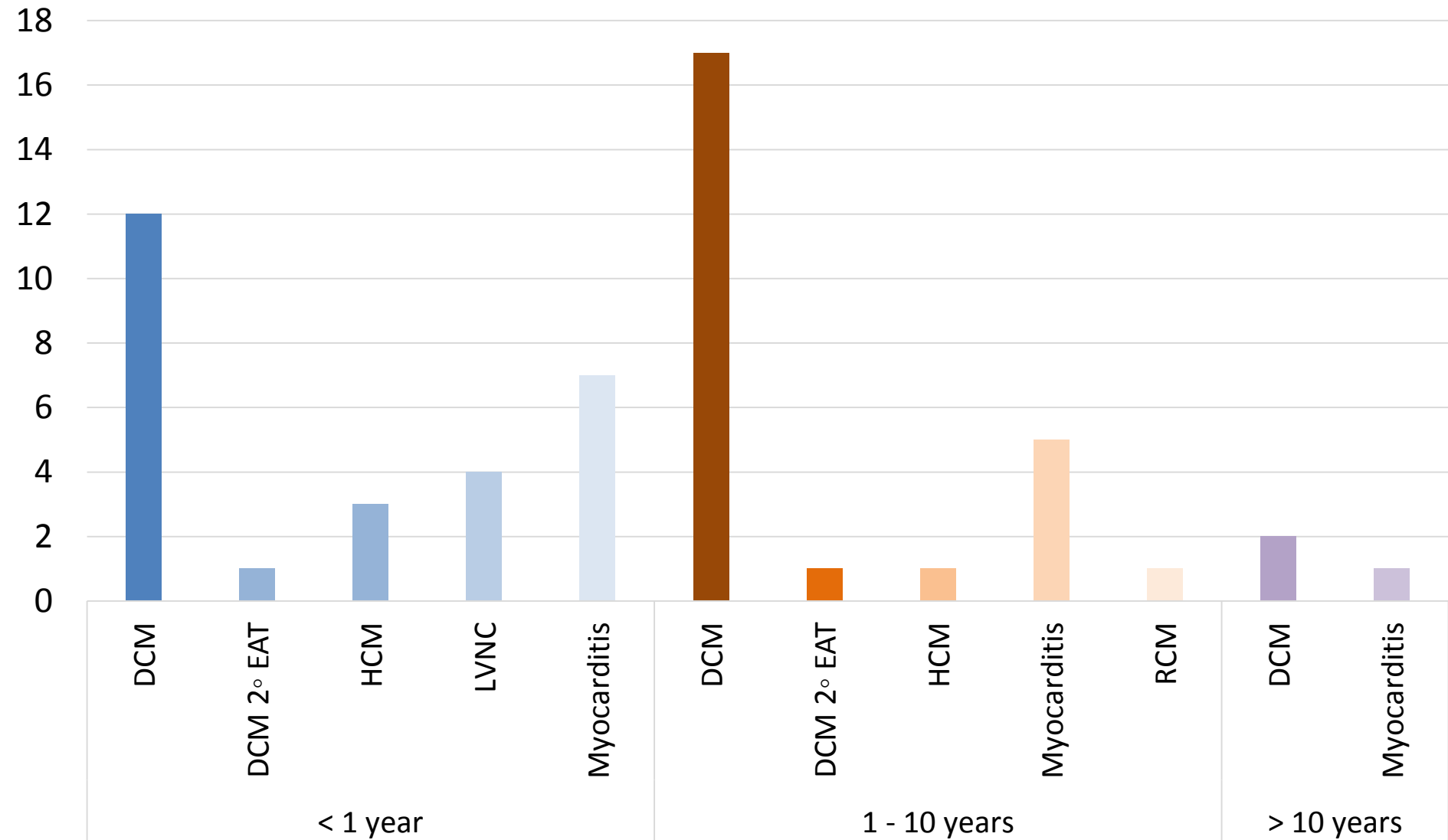
HIV exposure



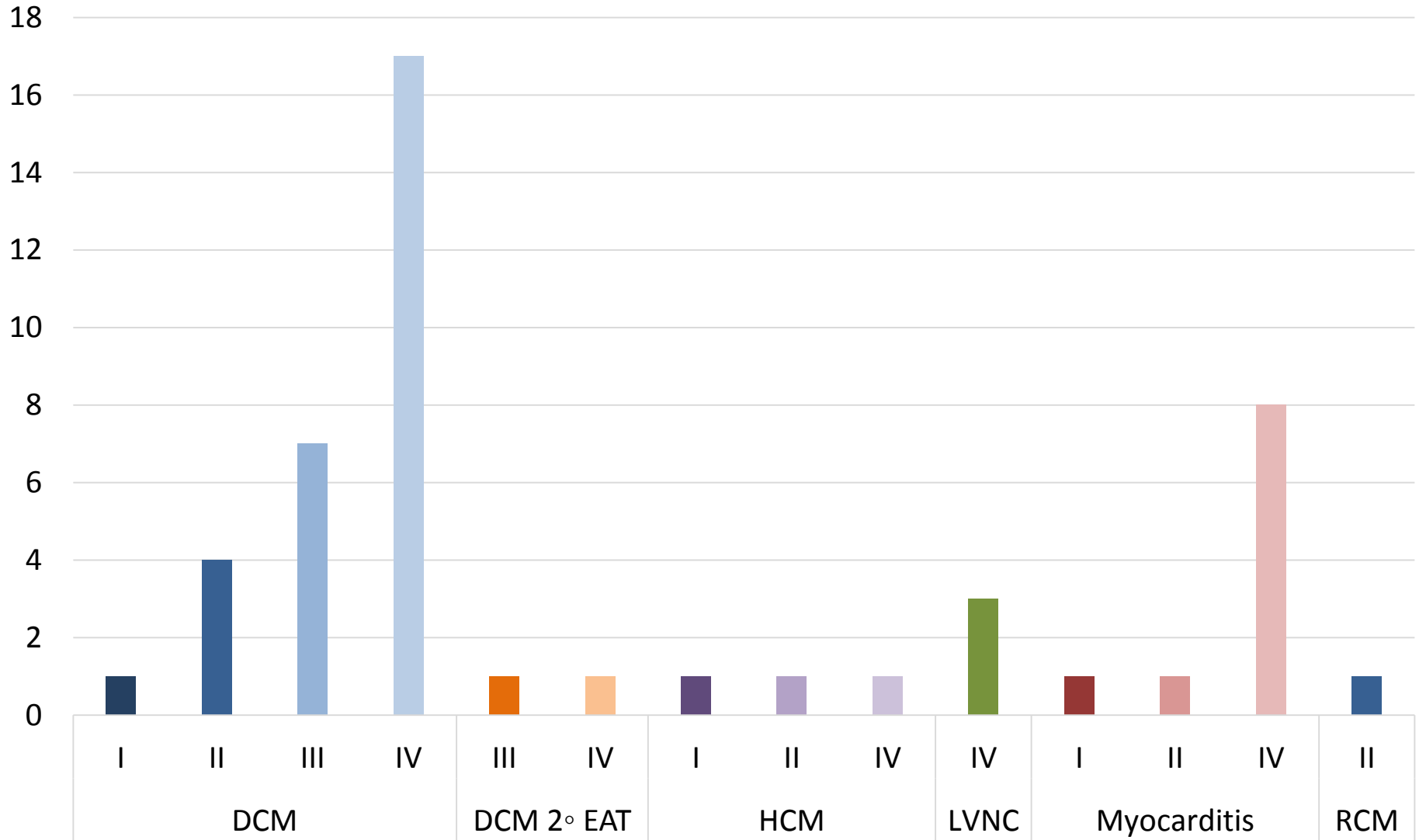
Diagnosis distribution



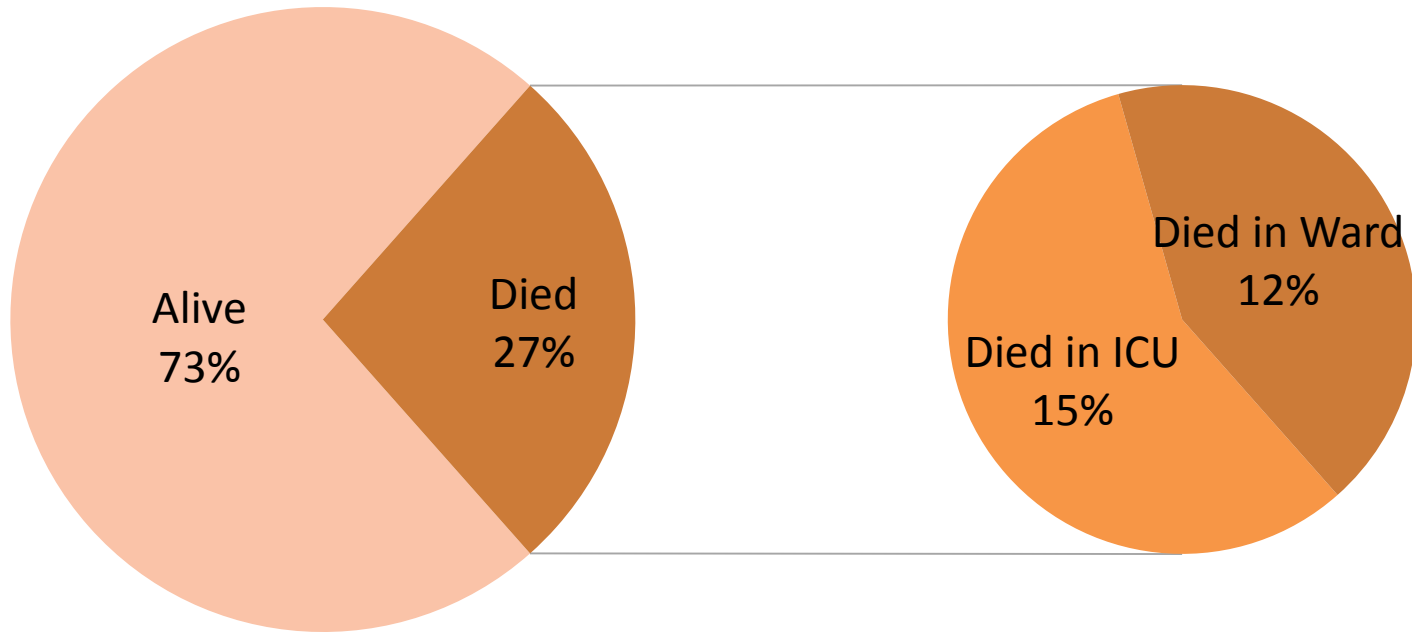
Diagnosis by age category



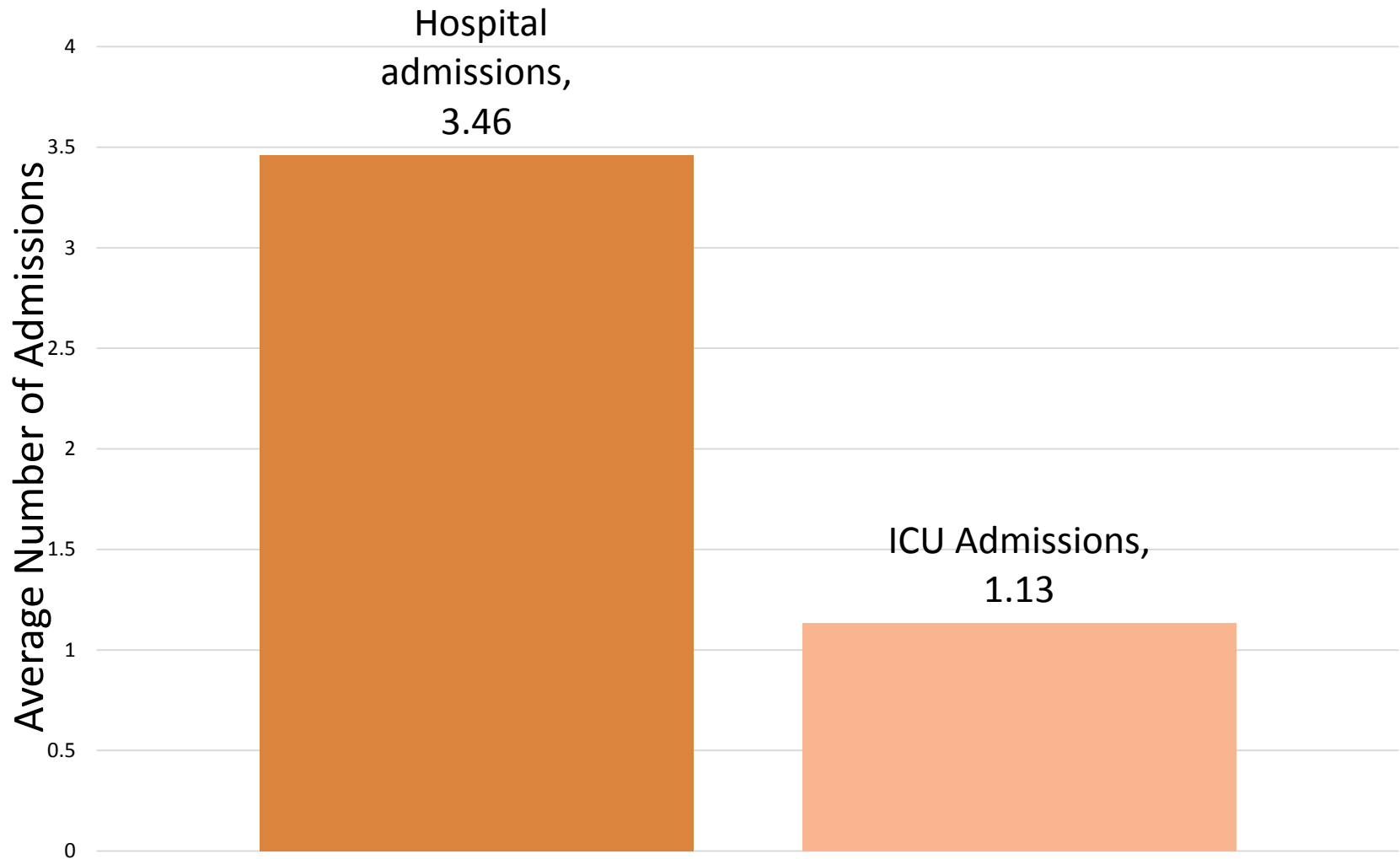
Ross classification by diagnosis



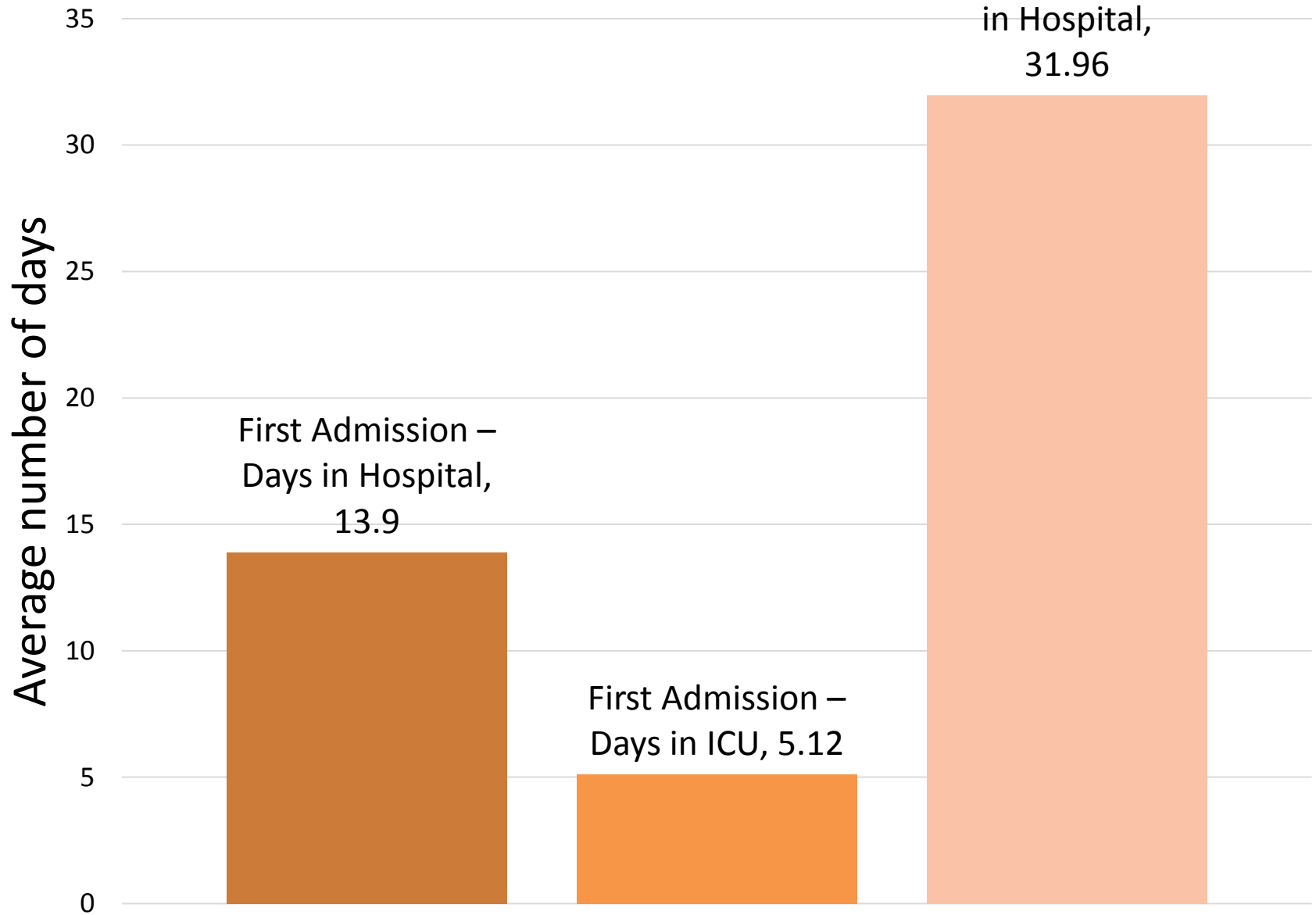
Mortality



Admissions to hospital and ICU (average)



Days in hospital/ICU on first admission and in total



Conclusions

- Large proportion of patients are infants / young children weighing less than 11 kg
- High incidence of ICU admission and multiple, prolonged hospitalizations – resource-intensive (beds, economic)
- IMHOTEP is proving to be an invaluable tool in the study of CMO in African children – first time systematic data
- Ultimate aim: improve prevention and management of cardiomyopathy / myocarditis
- Potential to build capacity locally, nationally and internationally in:
 - Echocardiography techniques/modalities
 - CMR: catalyst to initiating paediatric CMR program
 - Mechanical circulatory support (VADs) and heart transplantation

Acknowledgements

- Thomas Aldersley
- Barend Fourie
- Yanita Singh
- Sarah Kraus
- Liesl Zuhlke
- John Lawrenson
- Alexia Joachim





THANK YOU

