

Master thesis submitted to the Charité - Universitätsmedizin Berlin
in partial fulfilment of the requirements for the award of a
Master of Science degree in International Health

**Title: PAPUCO - the Pan African Pulmonary hypertension Cohort study:
Preliminary data analysis after one year of recruitment**

South Africa, 2012

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Key words: pulmonary hypertension, right heart failure, cardiovascular disease, HIV, tuberculosis, Africa

Abstract

Background: Pulmonary hypertension (PH) and consecutive right heart failure (RHF) is a "not-so-rare" form of heart disease of multiple aetiologies in Africa. PH is a devastating, progressive disease, with increasingly debilitating symptoms and, usually, shortened overall life expectancy. The epidemiology of PH in Africa and the distribution of its multitude of aetiologies has not yet been described, but limited reports suggest that the incidence of PH in Africa is higher than that reported from developed countries, owing to the pattern of diseases prevalent in the region. Many known risk factors for PH are hyperendemic in this part of the world, including HIV/AIDS, schistosomiasis, and hereditary haemoglobinopathies. A high prevalence of tuberculosis, poorly treated asthma, high levels of pollution in urban areas subsequently lead to various forms of pulmonary disease and PH. **Objectives:** To describe the demographics, socio-economic aspects, clinical presentation, aetiology and

outcome of PH in Africa. **Methods:** A prospective observational study, the Pan African Pulmonary hypertension Cohort study (PAPUCO) was established and launched in 2011. It is aimed to prospectively enrol patients with newly diagnosed PH who reside in Africa. The patients will be followed-up six monthly on an ongoing basis to establish a large Pan African patient cohort. Confirmation of PH will be by Echocardiography. **Results:** Among the 88 recruited cases, the median age was 38 years (range 1-86 years) with a female-to-male ratio of 1.6:1. Cardiovascular risk factors were family history of cardiovascular disease (34%), hypertension (28%), hypercholesterolemia (8%), diabetes (6%) and smoking (5%). The HIV prevalence of the cohort was 31% with a median CD4 count of 352 cells/ μ L (IQR 201-516) at presentation. Thirty percent of patients had previous episodes of tuberculosis (TB) with 13% more than one previous episode of TB. Presenting symptoms were shortness of breath (92%), fatigue (80%), palpitation (68%), cough (52%), cyanosis (17%) and syncope (or near syncope) (6%). The mean right ventricular systolic pressure was 56 mmHg (IQR 46-68), whereas the median RVSP in HIV-associated PH (HIV-PAH) was 60 mmHg (IQR 53-73, $p=0.08$). Final diagnosis was idiopathic PH (1%), HIV-PAH (13%), congenital heart disease PH (4%), PH due to left heart disease (52%), PH due to lung disease and hypoxia (20%), chronic thromboembolic PH (2%) and unclear and/or multifactorial mechanisms (8%). Of a sub-cohort with 22 HIV-PAH patients, 36% died within the first six months. The median time from diagnosis of PH to death was 3.0 months (IQR 1.5-3.7). The median RVSP at baseline was 72 mmHg (IQR 60-83) in patients who died within six months compared to a median RVSP of 55 mmHg (IQR 45-65) at baseline of patient alive at six month follow-up ($p=0.033$). **Conclusion:** Left heart disease, HIV, chronic lung disease and congenital heart disease are common contributors of PH in Africa. Disease targeted therapy is not available in the public sector and outcome of HIV-PAH is very poor. Further analysis of the PAPUCO cohort once outcome becomes available of all patients will give more insight on course of disease, outcome and predictors of outcome.