

ABSTRACT

Objective: To describe the demographics, nutritional status, clinical features and assess the outcomes of patients diagnosed with AOPA (anomalous origin of the pulmonary artery from the aorta) in a developing country.

Methods: Retrospective cross-sectional review of all children and adults with AOPA seen between April 1990 and April 2015 at the Chris Hani Baragwanath Academic Hospital, a tertiary care institution. All available clinical, radiographic, electrocardiographic, echocardiographic, computed tomography scan, cardiac catheterization and angiography, operative and follow-up data was reviewed from the case files and database.

Results: Seventeen patients (infants, n = 15) were diagnosed with AOPA and ten (59%) were male. The median age was 81 days (2 days-36 years). Two thirds (77%,10/13) of the patients were malnourished and 46% (6/13) had severe malnutrition. The most common presentation was congestive cardiac failure and severe pulmonary hypertension was present in all the patients. The diagnosis was made on echocardiography in ten patients (59%) and on catheterization angiography in six patients (35%). Fourteen patients (82%) had anomalous origin of the right pulmonary artery (AORPA). The most common associated cardiac defect was patent ductus arteriosus (PDA). Surgery was undertaken in three patients and three patients were deemed inoperable. The overall mortality was 82% (14/17).

Conclusion: High morbidity and mortality is associated with AOPA if early diagnosis and surgery is not achieved. Mortality is related to early development of pulmonary hypertension and progressive congestive cardiac failure. Late presentation and lack of adequate resources may be contributing factors to the mortality in a developing country.