Background: Pulmonary arterial hypertension (PAH) is commonly associated with congenital heart disease and relates to type of the underlying cardiac defects and repair history.

Objective: To describe the clinical and echocardiographic findings of a group of adult patients with pulmonary hypertension secondary to congenital heart disease.

Methods: Descriptive cross-sectional study with a retrospective collection of data obtained from clinical history and paraclinical reports.

Results: We included 74 patients with a median age of 31.5 years, of which 52 were women (70.2%). The main heart disease was interventricular septal defect, followed by patent ductus arteriosus. At least 2 defects were found in 32 patients (43.2%). Only 22 patients (29.7%) had history of previous intervention and the predominant classification was Eisenmenger Syndrome. Most were found in functional class I-II. 86.5% (64 patients) received pulmonary vasodilators medications. Polycllobula was documented in more than half of the patients, with a high percentage of iron deficiency and thrombocytopenia. In the right catheterization, the average pulmonary artery pressure was 60 ± 22 mmHg, with a median pulmonary vascular resistance of 8.4 UW. In echocardiography, the main alteration was dilatation and hypertrophy of the right ventricle, with dysfunction in less than half of the cases. The predominant valvular disease was tricuspid regurgitation, followed by pulmonary insufficiency. The size and function of the left ventricle were normal in the majority.

Conclusion: Pulmonary hypertension affects a large group of patients with congenital heart disease, which have clinical and echocardiographic characteristics that differ from other groups of pulmonary hypertension.