

Clinical Presentation, Therapy and Prognosis of Pulmonary Arterial Hypertension in Patients with Congenital Heart Defects: An Analysis Based on the Data of the German National Register for Congenital Heart Defects

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Background:

5-10% of patients with CHD develop PAH. We provide an overview over the spectrum of disease, clinical presentation, therapy and outcome based on the data of the German National CHD Register.

Patients and Methods:

We included patients >1 year of age with persistent PAH after shunt closure or unoperated/palliated patients (including Eisenmenger syndrome [ES]).

Patients with isolated postcapillary pulmonary hypertension, patients in whom pulmonary pressures normalized after timely surgical intervention, patients with idiopathic PAH or patients with persistent PAH of the newborn were excluded. We defined the presence of PAH in accordance with current guidelines. In the absence of recent right heart catheterization data an estimated systolic pulmonary pressure >40mmHg measured on echocardiography was also accepted for the diagnosis. We collected data on clinical symptoms, exercise limitation, medical therapy and outcome. We included 184 patients (mean age 24.6±14.9 years, 58.7% females). Of these, 108 patients had ES. Eisenmenger patients were approximately 10 years older than the remaining patients (mean age 29.3±12.8 years vs. 17.8±15.2 years, p<0.001). Overall, 61.9% of patients were in NYHA class III. The mean 6-minute walk test distance was 382±122 m (ES 368±118 m). Overall, 44% of patients (ES 51%) received advanced PAH specific therapies (69.9% Bosentan, 25.3% Sildenafil), 14% received dual medical therapy. 51% of patients received heart failure medication. 16% of patients were treated with oral anticoagulants (ES 15%), while 22% (ES 25%) of patients received Aspirin. The mean survival rate at 1, 3, 5 and 10 years of follow-up was 94%, 83%, 77% and 64% in the entire cohort, whereas survival was even worse in ES patients (93%, 76%, 66% and 49% respectively).

Conclusions:

Despite the availability of widespread and timely surgical correction for CHD patients in Germany, we could identify a considerable number of PAH-CHD and especially ES patients. The majority of patients are symptomatic and have a reduced exercise capacity. A large proportion of patients received heart failure medication and especially Aspirin, which is not supported by current recommendations. Additionally, our data illustrate the poor prognosis of PAH-CHD patients despite the use of disease targeting therapies in 44 % of patients.