Pulmonary arterial hypertension is a rare life threatening disease affecting relatively young people. From the early stages of disease patients show a reduced exercise tolerance with a characteristic cardio-pulmonary exercise profile. Research shows that assessment of functional status with measurement of heart rate and gas exchange is clinically useful. However, like in all areas of science there are still many questions to answer: Do the more comprehensive exercise tests provide the most relevant information? Which parameters should we use? Will these patients benefit from training? Is the cardio-pulmonary exercise profile influenced by the cause of the disease? We tried to answer and discuss most of these questions based on original research in the chapters of this thesis.

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