Respiratory Care 2013 Jul;58(7):1246-54. doi: 10.4187/respcare.02252. Epub 2012 Dec 27.

Complications leading to sudden cardiac death in pulmonary arterial hypertension.

Demerouti EA1, **Manginas AN**, Athanassopoulos GD, Karatasakis GT.

Abstract

Pulmonary arterial hypertension (PAH) is a disease of small pulmonary arteries, characterized by vascular proliferation and remodeling. Progressive increase in pulmonary vascular resistance ultimately leads to right ventricular heart failure and death. PAH-specific drug therapy has improved clinical outcomes and survival. While the survival is better, progression of pulmonary vasculopathy contributes to pulmonary artery dilatation. Left main compression syndrome, pulmonary artery dissection, pulmonary artery rupture, and severe hemoptysis are reported as complications leading to sudden cardiac death, an event encountered more often in PAH patients. The advent of PAH-targeted drug therapy has reduced referral for lung transplantation; however, severe complications require rapid diagnosis, decision making, and possible registration on a lung transplantation waiting list. PAH referral centers provide multidisciplinary emergency care and specific therapeutic management, contributing to improved quality of life and survival for PAH patients. We review the complications leading to sudden death in PAH.

KEYWORDS:

left main compression syndrome; pulmonary artery dissection; pulmonary artery rupture; pulmonary hypertension

PMID: 23271814 [PubMed - indexed for MEDLINE]