

Clinical Characteristics

- Increased right atrial pressure [16]
- Decreased cardiac index [16]
- Increased mean pulmonary artery pressure [9,12,15,17]
- Increased pulmonary vascular resistance [12,18,19]
- Difficulties during pregnancy [19]
- Right sided-heart failure [6,8,9,12,17,19,20]
- Morbidity/mortality [6,15,17,19-21]
- Increased risk of infection [19]

Symptoms

- Shortness of breath/ dyspnoea/ breathlessness [6,8,12,15,21,23,24]
- Chest pain [6,12,24]
- Palpitations [12,24]
- Syncope/Dizziness [6,12,24,25]
- Tiredness/Fatigue [6,8,9,12,24]
- Peripheral oedema [6,9,12,24]
- Cough [12]
- Cyanosis [12,24,25]
- Feelings of weakness
- Headache

Functional Impairment

- Reduced exercise capacity [9,14-16,19,21,22]
- Physical functioning [6,8,9,12]
- Activities of daily living [8,14]
- Travel [19]
- Problems with sleep [8,9]
- Social impact [6-9,12,21]
- Psychological functioning [6-8]
- Emotional impact – depression/ anxiety [6,8,9,12,21,23]
- Role functioning [7]
- Control of own life
- Cognitive functioning
- Work

Treatment options

- Vasodilator therapy [6,15,17,19,21]
- Calcium channel blockers [19,20]
- Anticoagulant drugs [15,19-21]
- Supplementary oxygen [21]
- Transplantation [12,17,19,20]
- Diuretics [15,19,21]
- Prostaglandins [12]
- Endothelin receptor antagonists [15,16,21,25]
- Prostanoids [26]
- Phosphodiesterase type-5 inhibitors [19,20,25,26]
- Antiproliferative agents [21]
- Cardiac glycosides [22]
- Balloon atrial septostomy [19]
- Exercise [21]

→ Causal factor
 - - - → Improving factor

