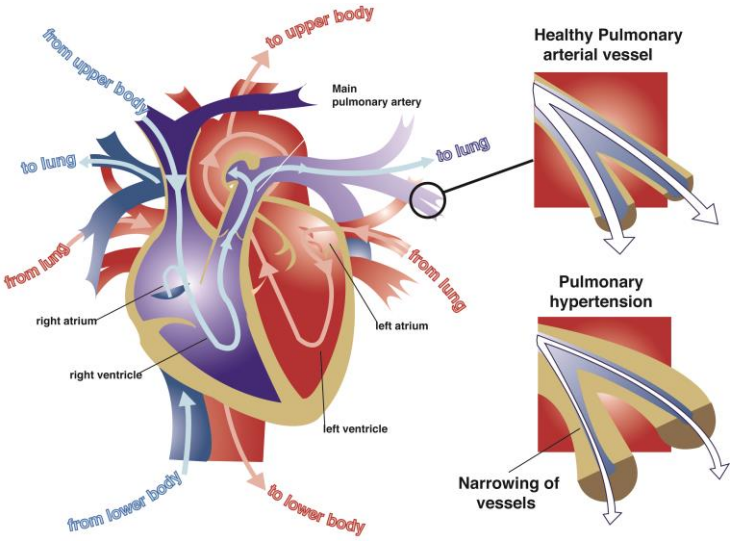
Autoimmune Resource and Research Centre
Information Sheet

Managing Pulmonary Artery Hypertension (PAH)

What is PAH?

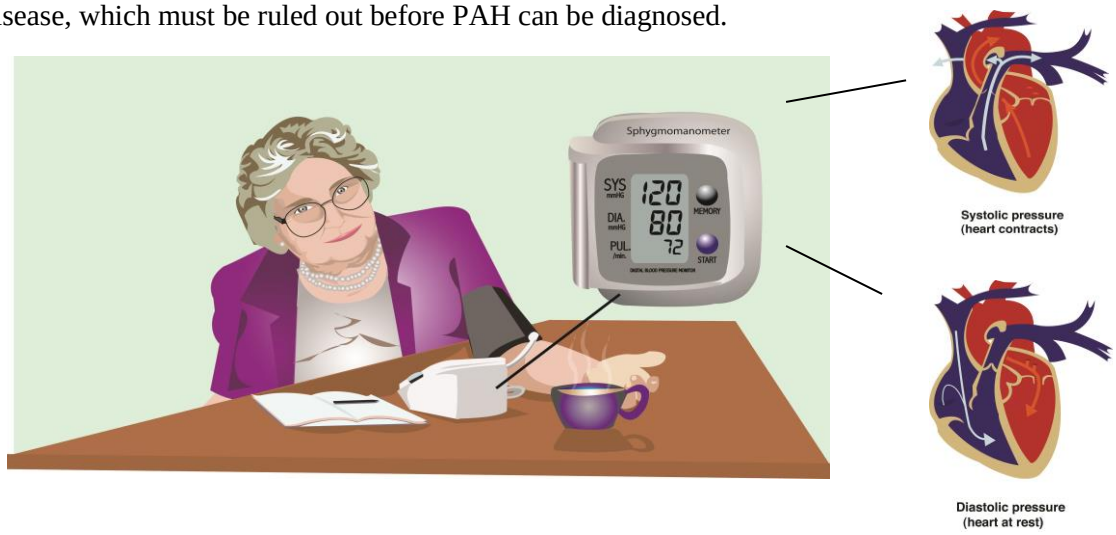
Pulmonary artery hypertension (PAH) is diagnosed when medical tests reveal increased resistance to flow due to narrowing of the vessels running from the (right-side of the) heart to the lungs.



Blood goes to the lungs to pick up oxygen before being pumped through the body by the left-side of the heart, so PAH often presents with shortness of breath due to reduced oxygen levels in the blood. The normal number for pulmonary vessel resistance (PVR) is less than 3 ("Woods") units, and pulmonary artery pressure (PAP) is normally less than 15 (mm Hg). Values for PVR of more than 3, or PAP more than 25, are the defining feature of PAH (as long as left heart function is normal). Because the right-side of the heart is pumping against a greater load, this strains the heart, causing palpitations, fatigue and chest discomfort, and may cause right-heart failure, which shows itself by breathlessness & dizziness on exertion and ankle swelling (oedema).

What PAH Is Not

PAH is different to what is commonly referred to as "blood pressure"(elevated systemic blood pressure (SBP), or systemic hypertension (SHT)), which is a raised pressure in the vessels supplying blood to the body from the left-side of the heart through the aorta and its branches, as reflected by readings detected using the "blood pressure cuff". PAP can also go up in people with lung disease, hypoxia, thromboembolism (clots), and left heart muscle or valve disease, which must be ruled out before PAH can be diagnosed.



Do PAH Treatments Work?

The three major classes of PAH-specific therapies include prostacyclin-analogues (such as iv epoprostenol and inhaled Iloprost), endothelin-receptor blockers (also called endothelin-receptor antagonists, or "ERAs": including Bosentan and Ambrisentan), and phosphodiesterase-5 (PDE5)-inhibitors (such as sildenafil). Recent pooled studies of the use of these therapies in PAH revealed that they reduced death rates by around 40% with good side-effect profiles. The choice of treatment involves considerations including dosage-convenience (once or twice daily dosing preferable), potential drug interactions (e.g. sildenafil and nitrates), and risks of side-effects (e.g. liver function changes with ERAs). Traditional supportive therapies, including warfarin and diuretics are often appropriate, and selected circumstances may warrant treatment with calcium blockers, digoxin and oxygen. On average, PAH-specific treatments increase the 6MWD by 40m.

What Non-Drug Approaches May Be Useful?

Pooled studies of specialised exercise programs in PAH reveal improvements of at least 90m in 6MWD. These results represent over twice the benefit seen with drug therapies, and suggest that supervised exercise programs should be a universal part of PAH management.

Evidence suggests that management within a specialised multidisciplinary PAH clinic (incorporating expertise of nursing, allied health, cardiology, respiratory, rheumatology and immunology specialists) also improves patient outcomes and satisfaction

The impact of stress reduction and positive attitudes in chronic medical conditions is also well-recognised, with encouragement, reassurance, regular communication, and provision of accurate balanced up-to-date knowledge (avoiding pessimistic messages) being critical.

Are Support Groups Helpful?

Support groups provide access to many of the helpful factors mentioned above, and are increasingly becoming available. Current clinical, support, educational and research activities undertaken by Hunter Health in the field of PAH are supported by Autoimmune ARRC in recognition of this close connection between autoimmunity and PAH. ARRC is a not for profit charitable organisation supported by medical trust funds, public donations and corporate endowments with the vision of providing world-best practice care, support, information, and research opportunities to individuals with systemic and organ-specific autoimmune conditions.

Pulmonary Hypertension Australia and Scleroderma Australia also provide valuable advocacy, educational and support roles.

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