Pulmonary hypertension (PHT), usually a late-stage complication, is the most common cause of death in mixed connective tissue disease (MCTD), occurring in up to 38% of patients. Since PHT does not become clinically manifest until advanced, even mild elevations in pulmonary arterial pressure reflect diffuse and extensive vascular damage. Through the mid-1980s median survival from the date of diagnosis was approximately 2.5 years. However, several novel therapeutic agents developed in recent years, have made early detection and monitoring imperative for improving long-term prognosis. ECHO with Doppler ultrasound is the common screening tool and should be performed to establish a baseline at initial diagnosis of MCTD. Pulmonary function testing (PFT) including diffusing capacity of the lung for carbon monoxide (DLCO) is a necessary part of the evaluation of all patients, primarily to exclude or characterize the contribution of underlying airway or parenchymal lung disease. A fall in DLCO responsive to medical management. Other agents that are currently being evaluated include phosphodiesterase inhibitors such as dipyridamole and sildenafil, inhaled nitrous oxide and arginine supplementation. Surgical techniques including lung transplant remain the mainstay of treatment for patients with PHT who are unresponsive to medical management.

References

4. Kim H, Yung GL, Marshall JH, Konopka RG, Pedersen CA, Chiles PG. Endothelin mediates pulmonary vascular remodelling in a canine model of chronic embo showing improved long-term survival with the use of calcium channel blockers. Prostacyclin derivatives (epoprostenol (IV), treprostinil (subcutaneous), iloprost (inhaled), beraprost (oral)) are potent vasodilators with antiplatelet aggregatory effects that have been shown to primarily improve exercise tolerance and survival to a lesser extent. Endothelin antagonists (bosentan, sitaxsentan, ambrisentan) block endothelin-1, which is a potent vasoconstrictor and smooth-muscle mitogen that might contribute to the increase in vascular tone and the pulmonary vascular hypertrophy associated with PAH. Other agents that are currently being evaluated include phosphodiesterase inhibitors such as dipyridamole and sildenafil, inhaled nitrous oxide and arginine supplementation.

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