Recommendations for Patient Referral to a PH Specialist

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Pulmonary arterial hypertension (PAH) is a rare disease. Patients with suspected PAH should have their doctors perform diagnostic testing based on their suspicion for PAH. This will often include an echocardiogram to estimate how high their pulmonary artery pressure is and other lab tests and studies of the lungs. If the echocardiogram suggests PAH, or if PAH is confirmed by right heart catheterization, it is appropriate for the treating doctor to refer the patient to a doctor that regularly evaluates and treats PAH (PH specialist).

A PH specialist along with the entire PAH team has the most experience managing PAH and has expertise with all approved classes of PAH-specific drugs, such as calcium channel blockers (CCBs), prostacyclins, endothelin receptor antagonists, sGC stimulators and phosphodiesterase 5 inhibitors. The PAH team may also be participating in clinical research trials that offer new, alternative investigational PAH therapies. Because all of these therapies require expertise in PAH, it is recommended that only PH specialty treatment centers initiate these treatments.

The follow-up care of patients receiving PAH-specific therapies should include both visits to the patient’s primary care physician or rheumatologist and periodic evaluations by the PH specialist. The frequency of visits should be individualized according to the patient’s specific clinical condition. It is also advisable to involve a local cardiologist or pulmonologist in the patient’s care. It is important to keep your PH specialist/nurse coordinator informed of changes in your treatment plan. This includes tests, medications and therapies your local health care team recommends. Coordination of care plays a big role in managing PAH patients.

Considerations for Children: Similar approaches have been recommended for infants and children with PH as well. At the time of initial PH diagnosis, a comprehensive history and physical examination in combination with diagnostic testing for assessment of PH etiology/classification and formal assessment of cardiac function should be performed prior to the initiation of therapy at an experienced center. Children with PH should be evaluated and treated in comprehensive, multidisciplinary clinics at specialized pediatric centers, with close communication with primary care providers and other medical centers. In general, outpatient follow-up visits at 3 - 6 month intervals are reasonable, with more frequent visits for patients with advanced disease or after initiation or changes in therapy.

