**FAMILIAL PRIMARY PULMONARY HYPERTENSION**

**HISTORY**
Familial Primary Pulmonary Hypertension (FPPH) describes a condition of markedly high blood pressure in the arteries of the lungs. This disease results in narrowing of the lumen of the pulmonary arteries. The resulting effect is the right side of the heart works harder than normal in an attempt to force enough blood to the lungs. The long term consequences are enlargement of the right side of the heart and increased cardiac stress which over time leads to heart failure.

**CAUSES**
FPPH is attributed to a genetic mutation on the BMPR2 gene located on chromosome 2. This mutation causes an over growth of cells in the small arteries of the lungs. Additional risk factors that increase the severity of FPPH include environmental factors such as diet, smoking, or the usage of certain diet medications.

**SYMPTOMS**
The most common symptoms of Familial Primary Pulmonary Hypertension include: shortness of breath, chest pain, weakness, fatigue, dizziness, swelling of the ankle or leg, and heart palpitations.

**TREATMENT**
There is no known cure for this disease so treatment focuses on controlling the symptoms related to FPPH. Lifestyle changes and medications are the mainstay therapies.

**TESTS AND DIAGNOSIS**
Diagnosis of FPPH begins with a complete patient history and physical examination. Questioning the patient will elucidate the most common symptoms. During the physical examination a physician may identify enlarged veins in the neck, swelling of the liver and spleen, and abnormal heart sounds. It is typical for a physical exam to reveal little during the early stages of this disease. It may take several months to make a definite diagnosis. Diagnostic tests include but are not limited to tissue biopsy, chest X-rays, CT scan, Echocardiogram (using sound), and pulmonary function tests.

**OTHER INTERESTING INFORMATION**
- Due to its genetic cause, this disease can not be prevented, however lifestyle and environmental changes can be made to alleviate some of the symptoms.
- The most severe complication of this disease is heart failure which may lead to death.
- Currently research is being done in the areas of exercise, respiratory therapy, and new medications to better treat the symptoms related to this disease.
- Approximately 1,000 new cases are diagnosed each year. The disease is twice as common in females than males.

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