

Idiopathic dilatation of the pulmonary artery (IDPA) is a rare congenital defect characterized by a wider than normal main pulmonary artery in the absence of any apparent anatomical or physiological cause. Idiopathic dilatation of the pulmonary artery commonly does not produce symptoms because there is no circulatory abnormality. Clinical signs are minimal, and usually consist of a palpable pulmonary ejection sound that disappears when the patient inhales, a soft pulmonary ejection systolic murmur (abnormal heart sound), and splitting of the second sound on breathing in. IDPA does not cause pulmonary valve disease, nor does bacterial endocarditis occur in patients with this condition. The electrocardiogram is normal, and diagnosis is made when chest X-rays reveal a dilated pulmonary artery without cardiac chamber enlargement. The cause of idiopathic dilatation of the pulmonary artery is unknown. A defect in the normal development of pulmonary artery elastic tissue before or after birth has been postulated. The dilatation may also be a consequence of a generalized connective tissue disease as it is occasionally found in Marfan's syndrome or Ehlers-Danlos syndrome. (For more information on these disorders, choose "Marfan" and Ehlers-Danlos" as your search terms in the Rare Disease Database. The incidence and prevalence of IDPA are not known. Because the disorder is benign in most instances, neither clinicians nor epidemiologists are able to measure the distribution of the disease with confidence. Treatment for idiopathic dilatation of the pulmonary artery is not required. People with this condition have a normal life expectancy, provided they have no cardiac lesions.