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Changing natural history of pulmonary hypertension in the last 20 years

O. Sitbon reviewed the Natural History of Pulmonary Hypertension during the last 20 years. He reminded us of the new classification of pulmonary hypertension with the main groups divided in: pulmonary artery hypertension; pulmonary hypertension related to left heart disease; pulmonary hypertension with lung diseases / hypoxemia or pulmonary hypertension due to chronic thrombotic or embolic diseases. Among the pulmonary artery hypertension the French collected in order of frequency, the idiopathic, the one related to connective tissue diseases, the one connected to congenital heart diseases and those related to portal hypertension are followed by the one secondary to appetite suppressant exposure and HIV infection. The familial forms are reported less frequently (about 5 %). For the evaluation of most cases, it is shown that during their progression, the pulmonary pressure tends to plateau while the pulmonary vascular resistance rises with a progressive decrease in cardiac output during exercise and later even at rest. The prognosis is best for congenital heart diseases and worst for pulmonary hypertension related to connective tissue diseases or HIV infection, being of 37 % and 21 % respectively at three years without treatment. These prognosis are being markedly improved with the new available treatment. It is recalled that idiopathic pulmonary artery hypertension (IPAH) is related to the gravity of dyspnoea with 58 months median survival for dyspnoea NYHA class I and II; 31 months for NYHA class III and 6 months for NYHA class IV. A distance above or below 250 m during the six-minute-walking-test is shown to influence the rate of survival by more than 25 % at 5 years even with I.V. Epoprostenol, the first treatment shown to change the prognosis of these patients. It was shown that the earlier the treatment with Epoprostenol, the better the prognosis. The patients with NYHA III and IV remain with a poor outcome with this monotherapy. New treatments are changing survival of IPAH, among those Bosentan and Treprostinil, being alone shown to be as effective as Epoprostenol in this respect. Thus, these new treatments appear to have completely changed the history of pulmonary hypertension during the past few years, decreasing the need of transplantations.

L. Nicod

Chairman of the session "Epidemiology and genetics of pulmonary hypertension"