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The Medical Bulletin

Clinical Case

What is hepatopulmonary syndrome?

Hepatopulmonary syndrome is defined as a clinical disorder associated with advanced liver disease, pulmonary vascular dilatation and a defect in the oxygenation of blood, in the absence of detectable primary cardiopulmonary disease.

The pulmonary vascular dilatations are caused by increased estrogen levels in the blood of patients with chronic liver disease (due to inability of the diseased liver to detoxify estrogen)

These vascular dilatations occur predominantly in the bases of the lung, and are sites of intrapulmonary arteriovenous shunting.

Therefore, when the patient is sitting up, blood pools at the bases of the lung with resultant increased intrapulmonary arteriovenous shunting.

This worsens the hypoxia and produces platypnea (shortness of breath relieved by lying down) and orthodeoxia (fall in the arterial PO2 in the upright position).

This hypoxia is responsible for the cyanosis and clubbing that occur in certain cases of chronic liver disease. Progressive and severe hypoxemia is an indication for liver transplantation and this is currently the only effective treatment for this condition.

Features of hepatopulmonary syndrome

- Advanced chronic liver disease
- Absence of primary cardiopulmonary disease
- Intrapulmonary vascular dilatation
- Intrapulmonary arteriovenous shunting Arterial hypoxemia.

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