ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Hepatopulmonary Syndrome: A Liver – Induced Lung Vascular Disorder.

Subhashini K*.

Department of Child Health Nursing, Sree Balaji College of Nursing, Affiliated to Bharath University, Chrompet, Chennai, Tamil Nadu, India.

ABSTRACT

The hepatopulmonary syndrome (HPS) is characterized by a defect in arterial oxygenation included by pulmonary vascular dilatation in the setting of liver disease. Patient of all ages can be affected. The Hepatopulmonary syndrome has three components : liver disease, pulmonary vascular dilatation, and defect in oxygenation. If hypoxemia and dyspnea develop in these patients in the absence of known intrinsic cardiopulmonary disorder, the hepatopulmonary syndrome must be considered.

Keywords: Liver disease, cyanosis, clubbing, dyspnea, hepatopulmonary syndrome .

Corresponding author

ISSN: 0975-8585

INTRODUCTION

Hepatopulmonary syndrome is characterized by a defect in arterial oxygenation induced by pulmonary vascular dilatation in the setting of liver disease. It is a rare complication of liver disease of varied etiology and indicates a poor prognosis. If hypoxemia and dyspnea develop in these patients in the absence of known intrinsic cardiopulmonary disorder, the hepatopulmonary syndrome must be considered. Relationship between cirrhotic liver disease and lung was first described by Flukiger in 1884 in a woman with liver cirrhosis and digital clubbing. In the year 1997, Kennedy and Knudsen coined the term 'Hepatopulmonary Syndrome'. Clinical features include digital clubbing, cyanosis, spider neavi. The hepatopulmonary syndrome is characterized by platypnea and orthodeoxia [1-5].

Etiology

Patients with HPS may be asymptomatic. Etiology of this syndrome remains unknown. Most commonly accepted hypothesis postulates that there is in adequate synthesis or metabolism of pulmonary vasoactive substance by impaired liver, leading to functional vasodilatation of the pulmonary vasculature producing hypoxemia.

Pathophysiology

Mechanism is unknown.

But is thought to be due to increased liver production or decreased liver clearance of vasodilators, possibly involving nitric oxide.

↓

Dilation of the blood vessels causes over perfusion relative to ventilation

↓

Ventilation perfusion mismatch and hypoxemia

↓

There is an increased gradient between partial pressure of oxygen in the alveoli of lung and adjacent arteries while breathing room air.

↓

Late in cirrhosis,

- Develop high output failure
- Less time in capillaries per red blood cell
- Exacerbating the hypoxemia.

Clinical Manifestation

- ✓ Dyspnea
- ✓ Digital clubbing
- ✓ Cyanosis
- ✓ Severe hypoxemia with orthodeoxia
- ✓ portal hypertension
- ✓ Esophageal varices

Diagnosis

Three imaging techniques can be used to confirm the presence of intrapulmonary vascular dilatation.

- Contrast – enhanced Echocardiography
- Perfusion lung scanning
- Pulmonary Angiography

Treatment

Medical and Symptomatic Therapy.Medical therapy for HPS has been disappointing

- ✓ Trails of various therapeutic options
- ✓ Steroids
- ✓ Non Steroidal anti inflammatory drugs
- ✓ Plasma exchange
- ✓ Chemotherapeutic agent
- ✓ Prostaglandin inhibitors
- ✓ O2 supplementation (2 to 4 l/min)
- ✓ Liver transplantation.

CONCLUSION

The hepatopulmonary syndrome is under recognized complication of chronic liver disease. It must be considered in every patient with advanced liver disease manifesting symptoms of dyspnea and hypoxemia, severe clubbing and cyanosis when the intrinsic cardiopulmonary disorders were ruled out.

REFERENCES

[1] Rodríguez-Roisin R, Krowka MJ. Hepatopulmonary syndrome–a liverinduced lung vascular disorder. N Engl J Med 2008;358:2378-87.

[2] G Agrawal, N Kumar, D Rosha. Hepatopulmonary Syndrome.Journal of Association of Physicians of India 2008;56:265-7.

[3] Fingerendphalangen ohne chronische Veränderungen an den Lungen oder am Herzen. Wien Med Wnschr 1884;34:1457.

[4] J.D. Collier,G. Webster. Liver and biliary tract disease. In: Nicki R.Colledge, Brian R. Walker, Stuart H. Ralston, editors.Davidson's st Principles & Practice of Medicine, 21 ed. Edinburgh: Churchill Livingstone;2010.p.973.

[5] MarcGhany, JayH.Hoofnagle. Approach to the Patient with Liver Disease. In:Dan L.Longo, AnthonyS.Fauci, DennisL.Kasper, StephenL.Hauser, J.Larry Jameson, Joseph Loscalzo, editors. Harrison's Principles of Internal Medicine 18th edn. USA:McGraw Hill;2012.p.2524.