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Hepato Pulmonary Syndrome in Children

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Abstract
Background: *Hepato pulmonary syndrome is an important problem in children with cirrhosis of liver. To evaluate incidence, clinical and investigational profile of hepato pulmonary syndrome in children, the following study was performed in pediatric cardio gastroenterology ward of PGIMER Chandigarh.*
Keywords: *Hepato pulmonary syndrome, Children.*

Introduction

Hepato pulmonary syndrome is an important complication of liver disease characterised by intrapulmonary vascular dilatation and hypoxemia.(1)

Prevalence of hepato pulmonary syndrome in adults with cirrhosis ranges from 4% to 29%. However, prevalence of hepato pulmonary syndrome and its outcome in children is unknown.(2)

Hepato pulmonary syndrome is defined as a triad of (a) Liver disease with or without portal hypertension, (b) abnormality in gas exchange (PaO_2 < 80 mm Hg) and alveolar – arterial difference > 15 mm Hg (c) Pulmonary capillary Vascular dilatation. Some studies have shown that it is most associated with liver cirrhosis and portac hypertension (8% – 17%).(3)

Symptoms of hepato pulmonary syndrome are divided into hepatic and hypoxic. Hepatic syndromes include icterus, hepatomegaly, ascites, splenomegaly, telangiectasia, collateral, circulation, edema of extremities. Hypoxic symptoms are dyspnea on exertion, platypnea, digital clubbing and cyanosis.(4)

Dyspnea is described as cardiac symptom of hepato pulmonary syndrome. It is gradual in onset. It is first symptom reported by patients. Presence of platypnea is suggestive of diagnosis of hepato pulmonary syndrome.(5)

Digital clubbing as a physical sign of hypoxemia is seen in this patient. However clubbing is reported as indicator of advanced disease.(1)

The duration of respiratory symptoms up to diagnosis of hepato pulmonary syndrome could be 4.8 + 2.5 yrs.(6)

Routine determination of oxygen saturation in room air via pulse oxymetry is an effective test to determine hypoxemia in patients with chronic liver disease and hypato pulmonary syndrome. (Saturation) <94.10 is considered abnormal). These patients need liver transplantation. However pulse oxymetry that normal values does not rule out hepato pulmonary syndrome.(7)

Orthodeoxia test is done in hepato pulmonary syndrome orthodeoxia is characterized by decrease in PaO_2 >4 mm Hg or to 5% from supine decubitus position up to standing position. (4)

Arterial blood gas shows hypoxemia, respiratory alkalosis. It is due to hyperventilation intended to compensate for hypoxemia.

Alveolar – arterial PaO_2 difference > 15 mm Hg is an early marker of hepato pulmonary syndrome.(4)

Based on these findings, hyperoxia test is indicated. It is carried out with measurement of arterial blood gases of patient while breathing room air.

Blood gases are subsequently measured after patient breathes oxygen at 100% for 10 minutes.

Hyperoxia rest is positive when PaO_2 is maintained < 150 mm 1tg. It indicates presence of vascular shunt.[8]

Methods

Fifty children aged 3 – 12 years who attended pediatric gastroenterology OPD of PGIMER Chandigarh from July 1993 to June 2003 with liver cirrhosis and dyspnea were admitted. History (prenatal history, onset of liver disease, dyspnea, progress of liver disease, family / sibling history etc), clinical features (e.g. Ascites, jaundice, GI bleed, peripheral edema, dyspnea, cyanosis etc), investigational profile (i.e. pulse oxymetry, hyperoxia test, USG whole abdomen, LFT, PT, Serum cerulospasmin, HBs Ag, HIV Serology, Auto immune marker, Torch profile, MRCP etc) were noted.

Results

Out of 50 cirrhotic of cirrhosis 10 had hepato pulmonary syndrome characterised by hypoxia, orthodeoxia, cyanosis, platypnea. Mean age of children (aged 3 – 12 years) was 6 years. Male was 30, female 20. Out of 50 cirrhosis children, 30 had extra hepatic biliary atresia, 10 had TORCH sero positivity, 1 had congenital hepatic fibrosis, 5 had wilson's disease, 3 had auto immune hepatitis, 1 had HBs Ag positivity. Out of 10 children of Hepato pulmonary syndrome, 6 had mild grade (PaO_2 80 – 90 mm Hg), 3 had moderate grade (PaO_2 60 – 80 mm Hg), 1 had severe grade (< 60 mm Hg) cyanosis, orthodeoxia, platypnea were present in all cases. All 10 children showed pulmonary vascular dilatation, arterio venues shunting on contrast echo cardiography / Technetium 99 labeled macro aggregated albumin scan.

However 8 children had HPS type I (diffusel) and 2 had HPS type II (focal fistula) variety. All 10 children had alveolar – arterial PaO_2 difference > 15 mm Hg. All 10 children were enrolled for liver transplant as priority basis.

Discussion

Hepato pulmonary syndrome is characterized by hypoxemia with presence of alveolar arterial difference > 15 mm Hg. Depending upon alveolar arterial difference there is classification of according to severity : - mild (with PaO_2 > 80 mm Hg), moderate (PaO_2 60 – 80 mm Hg). Severe (PaO_2 50 – 60 mm Hg), very severe (PaO_2 < 50 mm Hg).

Based on these criteria, patient is classified with severe hepato pulmonary syndrome (PaO_2 59 mm).

This classification is carried out to identify liver transplant candidate with poorest prognosis. Post transplant mortality increases with PaO_2 < 50 mm Hg.[4]

Chest X-ray is normal in patients with hepato pulmonary syndrome. CT of the chest is indicated to exclude abnormality in pulmonary parenchyma causing hypoxemia.[9]

The presence of vascular dilatation can be seen. Functional spirometry and pulmonary volume study tend to be normal or mildly abnormal according to lung restrictions.[4]

Demonstration of intra-pulmonary shunt by echo cardiography with agitated saline test is considered gold standard for diagnosis of hepato pulmonary syndrome. It has sensitivity of 68 – 75% and specificity of 93 – 100%. It is positive if injection of 10 ml agitated normal saline into systemic vein, micro bubbles appear in right atrium after 3 seconds.

Method for confirming hepato pulmonary syndrome by demonstration of pulmonary vascular dilatation is pulmonary perfusion scan by using albumin macro aggregates labeled with technetium – 99 (99 m Tg to MAA). This identifies quantification of percentage of intra pulmonary dilatation based on greater extra

pulmonary uptake of macro aggregates. It is considered pathologic when values are ≥ 6%.[10]

According to Krowha classification, hepato pulmonary syndrome is divided into two groups.

a) Type I (diffuse) – patients with pre-capillary dilatation who respond to 100% O2, achieving PaO_2 > 200 mm Hg.
b) Type II (Focal fistulas) – patients with localized vasodilatation, similar to arterio venious malformation who have a poor response to administration of 100% O_2. These patients present true shunts, who are unable to attain PaO_2 > 200 mm Hg. It is less common than type I.[8]

Out of 10 cases of Hepato Pulmonary Syndrome in our series, 8 had type I and 2 had type II variety.

There is no effective medical treatment for hepato pulmonary syndrome. Liver transplantation is only treatment which reverses hepato pulmonary syndrome. Its success rate is 70 – 80%. However hepato pulmonary syndrome that is severe grade (PaO_2 < 50 mm Hg) shows mortality rate > 30%. [11]

Resolution of pulmonary dilatation is observed from 3 months post transplantation.[5]

Home oxygen therapy is indicated for the patients who have PaO_2 <60 mm Hg. It prevents circulatory complications of hypoxemia and improves quality of life while waiting for transplant.[8]

Poor prognostic factor are PaO_2 < 50 mm Hg in room air, shunt >20% on scan with albumin macro aggregates, severe pulmonary hypertension, positive hyperoxia rest.[10]

Prognosis without transplant is very poor, with a survival rate of 16 – 30% at 1 year from time of diagnosis.

Conclusion

Hepato pulmonary syndrome should be searched in children with liver disease having dyspnea and hypoxemia who are in a liver transplantation protocol. Its diagnosis will get priorities in transplant waiting list.

Conflict of interest – Nil

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