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# Hepatopulmonary Syndrome
## A Study Of Six Patients In Iraq With Literature Review

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## *Abstract*

***Among the pulmonary syndromes associated with liver disease is the hepatopulmonary syndrome (HPS). This is characterized by the triad of liver disease, intrapulmonary vascular dilatations (IPVD), and arterial hypoxemia (1,2). We studied six patients on the bases of clinical grounds and the available investigations in our country. English-language on case studies and series reporting results from patients with the HPS were reviewed.***

***Keywords : hepatopulmonary syndrome***

## *Introduction*

The term hepatopulmonary syndrome (HPS) ,first suggested by Kennedy & Knudson in 1977(3), is a functional process similar to the functional renal failure widely known as the hepatorenal syndrome(4). Impaired arterial oxygenation is a hallmark of the syndrom(1).Mild hypoxemia is seen in approximately two thirds of all patients with chronic liver disease(5)..Severe hypoxemia is less common with cirrhosis alone and when obvious cardiopulmonary causes are discarded, it is attributed to HPS. Intrapulmonary vascular dilatations (IPVD) are thought to be the cardinal cause of severe hypoxemia and are the defining feature of the HPS (6).These vascular abnormalities contribute to the mismatch between ventilation and perfusion, and right to left blood flow shunting(7).Several features characterize this syndrome. Most patients will present with manifestations of liver disease.

Spider-nevi are common ) Pulmonary Features include digital clubbing,cyanosis, dyspnea (dyspnea in upright position and relieved and platypnea (dyspnea....) by recumbence)(8). Several radiographic abnormalities may be seen in the chest radiographs of cirrhotic patients but more specifically ,the HPS may be accompanied by characteristically increased basilar interstitial and pulmonary vascular markings(1). These vascular abnormalities can be confirmed by one of the following contrast - enhanced echocardiography, perfusion lung scaning and pulmonary arteriography .Pharmacologic therapies for the HPS have been disappointing .Regarding recent studies liver transplantation is the treatment of choice in patients with severe HPS(2).

The aim of this study is to report the HPS in IRAQI patients and to bring the attention of clinicians especially cardiologists for the

possibility of this syndrome before proceeding to the invasive procedure of cardiac cathterization in cyanotic patient.

## *Patients And Methods*

During a period starting in Feb.1997 to April 1998, We studied six patients referred to us suspected to have liver diseases. Allof them were initially managed by cardiologists and investigated (including cardiac catheterization in 4 patients) for the suspicion of intracardiac defects. Full history and detailed Clinical examination done for every patient and a study protocol was filled Investigations included were: full blood count, liver function tests, serum chemistries, serum protein electrophoresis, coagulation tests viral markers, immune markers, screening for Wilson disease, , alfa one Antitrypsyn estimation (A1AT),pulmonary function tests ,chest radiographs, electrocardiogrammes, echocardiography ,abdominal sonography, asitic fluid analysis, upper GI endoscopy and lastly liver Biopsy

One patient (case No.6)was a heavy alcoholic and presented with hematemesis. Emergency endoscopy and injection sclerotherapy at the same time were carried out for oozing grade four esophageal varices. Another patient(caseNo.5) has been appointed for cardiac catheterization ,but on consulting us for his jaundice, the procedure was canceled.

## *Results*

Clinical characteristics and results of important investigations of the patients are shown in table 1 and 2.

Four patients(No.1,2 5,and 6)had established liver cirrhosis, of which three of them were of unknown cause. The last patient was a young heavy alcoholic man, he was also HbsAg +ve but his liver biopsy showed features of alcoholic cirrhosis.

patients(No.3 and 4)had chronic active hepatitis.The patient with immune hepatitis (No.4) was kept on steroid and azathioprim. Her serum transaminases and gamma globulin were significantly improved. But there was a mild improvement in her dyspnea and platypnea. One alfa interferon was not available for patient No.3( antiHCV Ab +ve)despite laboratory and histological indications for this treatment.linical characteristics of 6 patients with HPS

Table :1clincal characferistics of patients with HPS.

| Case No | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|
| Age (y) /sex | 11 y /F | 14 y /M | 16 y / M | 18 y /F | 20 y /M | 25 y/ M |
| Platypnea | Moderate | Moderate | Moderate | Moderate | Marked | Marked |
| Spider naevi | Numerous | Numerous | Numerous | Numerous | Numerous | Numerous |
| Cyanosis | Deep | Deep | Deep | Deep | Moderate | Deep |
| Clubbing | Drumstick | Drumstick | Grade III | Drumstick | Drumstick | Grade III |
| Jaundice | Mild | mild | Moderate | Moderate | Mild | Deep |
| Ascites | Mild | mild | mild | Moderate | Mild | Moderate |
| Splenomegaly | 4 cm BCM | Just palpable | Just palpable | 6cm BCM | 4cm BCM | 6cm BCM |
| Others | - | | Bilateral gynecomastia | Leg edema + | - | Leg edema + Dupuytren's contracture, bilateral gynecomastia. |

Table 2Results of important investigations in 6 patients

| Case No. | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|
| Viral markers | Neg. | Neg. | Anti HCV Ab +ve | - | - | HbsAg +ve |
| Immune markers | Neg. | Neg. | Neg. | ANA & ASMA +ve. | Neg. | Neg. |
| Wilson screening | Neg. | Neg. | Neg. | Neg. | Neg. | Neg. |
| Alfa one A.T | Normal | Normal | Normal | Normal | Normal | Mildly |
| gamma-globulin level | Mild ↑ | Moderate ↑ | Giant ↑ | Moderate ↑ | Moderate ↑ | Moderate ↑ |
| Pulmonary F. Ts. | Normal | Normal | Normal | Normal | Normal | Normal |
| Echo cardiography (no contrast) | Normal | Normal | Normal | Normal | Normal | Normal |
| OGD | EV Grade 4 | EV Grade 3 | EV Grade 4 | EV Grade 3 | EV Grade 4 | EV Grade 4 |
| Chest radiographs | Bibasilar shadowing | Same | Same | Same | Same | Same plus mild L pleual effusion |
| Liver Sonography | In homogenous nodular | Inhomogeneous nodular | Inhomogeneous | Inhomogeneous | Inhomogeneous nodular | Inhomogeneous |
| Cardiac Cath. | No shunt Normal p.a.p. | No. shunt Low p.a.p. | No. shunt Low p.a.p. | No. shunt Low p.a.p. | Not done | Not done |
| Liver Biopsy | Mixed cirrhosis | Mixed cirrhosis | CAH | CAH | Mixed cirrhosis | Alcoholic cirrhosis |

ANA : Antinuclear antibody Pulmonarey F.Ts : Pulmonary Function tests ASMA : Anti smooth muscle antibody P.a.P : Pulmonary artery pressureAlfa one AT. : Alfa one Antitrypsin OGD: Oesophagogastroduodenoscopy EV:Esophageal-Varices.CAH:Chronic Hepatitis

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# کۆنیشانی جگەر سیی
# لێکۆڵینەوەی شەش نەخۆش لە عێراق دا و پێداچوونەوەیەکی مێرووی نەخۆشی یەکە

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## پوختە

لە نصوئەوکۆنیشانانەی سی داکە هاوەڵێی نە خۆشیەکانی جگەرئە کات کۆنیشانی جگەرسی یە.
ئەم نەخۆشی یەش لەم سص بینە یە گصك ها تووە؛نە خۆشی جگەر،فراوان بوونی لووڵە کانی خوصنی سی یەکان،لە ڤەڵ کە م بوونە وەی ژمارەی ئۆکسجینی ناو خوصن بەرەکان.بۆ یەکەم جار هە ستاین بە لصکۆلینە وەی شەش نەخۆش لە (مدینە الگب) لە ساڵانی (١٩٩٧–١٩٩٨ ) لەم لصکۆلینەوەیەدا گشت بەست بووین بە نیشانەکانی نە خۆشیە کەو ئەو گشکنینە تاقیگەییانەکە لە عصراق دا دەست دە کەون.گصداچوونە وەیەکیشمان کرد بەوبڵاوکراوە ئینگلیزیانەدا کە تاك تاك ئەم نەخۆشی یەیان تۆمارکردووە،هەروەها دەرئەنجامی لصکۆلینەوە فراوانەکانیش لە سەر ئەم کۆنیشانە.

# متلازمة الكبد الرئوي
# دراسة ستة مرضى في العراق مع مراجعة ماضي المرض

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## الخلاصة

الدموية داخل الرئتين مع نقص اوكسجييني شرياني .لقد قمنا ولأول مرة في العراق بدراسة ستةمرضى في مستشفى مدينة الطب١٩٩٧–١٩٩٨معتمداعلى العلامات السريرية والفحوصات المتوفرة في القطر.و تمت مراجعة المقالات المنشورة باللغة الأنكليزية حول تدوين الحالا ت المنفردة وكذلك استنتاجات البحوث الشاملة حول هذه المتلازمة.بين المتلازمات الرئوية التي تصاحب امراض الكبد هي متلازمةالكبدالرئوي. هذه المتلازمة تتكون من الثلاثية ؛ مرض الكبد،توسع الاوعية .

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