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Egyptian Hepatic Veno-Occlusive Disease: Surgical Point of View

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1. Introduction

Hepatic veno-occlusive disease (HVOD): was described as a non portal cirrhosis occurring frequently in children and occasionally in adults. Now it is considered an important cause of non cirrhotic portal hypertension particularly in children [1].

Rollins 1989 [2], stated that HVOD is a non-thrombotic obliteration of small intrahepatic veins by loose connective tissues. The venous occlusion may be progressive and lead to massive hepatocellular necrosis. However the precise pathogenesis is still obscure but also most likely relates to venous endothelial injury.

Originally the syndrome was described in South Africa at 1920, but at present it is endemic in Jamaica, encountered in Afghanistan and India. The syndrome was described under different names, from Jamaica the disease was described under the term Jamaican veno-occlusive disease, in India the disease was given the term Indian childhood Cirrhosis (ICC), in Europe HVOD has been called endophlebitis obliterans of which sporadic cases were described, as in Germany. Hepatic veno- occlusive disease was examined by scanning electron microscopy (SEM). SEM correlated its histology and postmortem examination and disclosed microscopic occlusion of the centrilobular and sublobular veins in the liver, these veins were occluded partially or completely by intimal and medial thickening of their walls due to proliferation of collagen and reticulin fibers. In addition to venous obliteration, which had not been demonstrated by other techniques, frequent occlusion of the sinusoidal opening into the central veins was observed by SEM. [4], [5], [6].

Causes of non cirrhotic portal hypertension	
Intrahepatic	**Extrahepatic**
Schistosomiasis	Extrahepatic portal vein thrombosis
Extrahepatic portal vein thrombosis	Splenic vein thrombosis
Biliary cirrhosis, primary and secondary	
Chronic veno-occlusive disease	
Chronic active hepatitis	
Congenital hepatic fibrosis	
Haemochromatosis	
Alcoholic fibrosis	
Sarcoidosis	
Nodular regenerative hyperplasia	
Idiopathic portal hypertension	
Non-cirrhotic portal fibrosis	

Hepatic veno-occlusive disease has been recognized as being due to the toxic effects of some remedies, recently pyrrolizidine alkaloids mostly involved, as in senecio (bush teas) and crotalaria (comfrey trees). It is also now seen as complication of high dose of anti-neoplastic chemotherapy, especially in the setting of bone marrow transplantation. HVOD may be familial, so the term "*veno occlusive familial hepatic disease*" [7], [8], [9].

2. Hepatic veno-occlusive disease (HVOD) in Egypt: Overview

In Egypt Hashem 1939 [7], gave the first reference to this syndrome, in his study of portal cirrhosis among Egyptian children. Since 1939 several reports pointed out the occurrence of a specific syndrome among Egyptian children who rapidly developed abdominal distention with ascites and hepatomegaly. In 1965, Safouh et al [11]; reported that 54 Egyptian children were studied and the term "Hepatic vein occlusion disease in Egyptian children" was applied. At the same year, El Gholmy 1956 [10], studied a group of patients and introduced the term "Infantile cirrhosis of Egypt"

The different reports from Egypt, thereafter, describing the syndrome, the clinical picture, the pathology and the etiology revealed that HVOD is not uncommon among Egyptian infants and young children. They also have shown clearly for the first time that hepatic vein occlusion should be considered in the diagnosis of Egyptian children presenting with hepatosplenomegaly [11].

Safouh 1965 [11], reported that the Egyptian hepatic vein occlusion is the result of enhanced thrombotic activity of the blood with the formation of fibrinous thrombi followed by organization and thickening or closure of the vessels, a finding which seems peculiar to the Egyptian cases and thus differs from the classical HVOD.

3. Clinical Picture of HVOD

Clinical diagnosis is based on; hepatomegaly and/or right upper quadrant pain, ascites or unexplained weight gain and also jaundice may or may not present [7].

The acute stage starts abruptly with abdominal discomfort or pain accompanied by hepatomegaly and ascites, nausea and vomiting are common. Histologically the liver shows an edematous endophlebitis of the central veins associated with centrilobular congestion, hemorrhage and necrosis. Mclean 1969 [12], has shown experimentally that the block occurs first at the outlets of the sinusoids. Patients surviving the acute stage may progress to the subacute stage with persistent hepatomegaly and ascites which then diminish if an adequate collateral circulation becomes established. The chronic stage is a centrilobular type of septal cirrhosis [7].

Clinical picture:
Non febrile onset
Mild continuous dragging pain in right hypochondrium
Anorexia, nausea and vomiting
Rapidly filling ascites
Distended veins over the abdomin
Oliguria and pedal edema
Hepatomegaly
Splenomegaly in some cases

Tandon 1977

4. Diagnosis of HVOD

In the acute phase the diagnosis is usually readily made from the history and the characteristic clinical picture. In the sub-acute and chronic stages the diagnosis may be more difficult. In all stages the diagnosis is confirmed by the characteristic histopathological findings of liver biopsy in the absence of extrahepatic venous obstruction.

4.1. Laboratory Studies:

1. Safouh et al; 1965 [11] reported the following results:

Most of the cases showed some degree of anemia.

Total and differential leukocytic counts did not show any constant deviation from normal.

Liver function tests showed that : * Serum bilirubin was always below 3 mg/dl, * Serum AST varied between 20 and 60 units, * Serum ALT and alkaline phosphatase were found to be normal.

Erythrocyte sedimentation rate (ESR) was low in spite of advanced state of the disease.

The pattern of serum total proteins showed a state of hypoproteinemia ranging from 4-5 gm/dl and the albumen fraction is usually is decreased but globulin fraction may be increased.

2. Millis and Bale 1976 [13], stated that a feature of their cases is the partial immune deficiency. However, such a state of hypogammaglobulinemia reported by them goes parallel with findings in the acute cases only, that their cases were quite a different group of patients suffering from genetic immunodeficiency as observed from the very early appearance of the syndrome in some of them being as early as days.

3. Serum procollagen type III is an early and sensitive marker in VOD after BM transplantation, usually above 100 ng /ml.

4. Serum protein S,C, liedin factor

4.2. Ultrasonographic scaning of the liver:

It is of *definite help* in the diagnosis of this syndrome, it showed that the liver is enlarged especially the caudate lobe, splenic enlargement is usually of mild degree and ascites is always found in acute cases. Narrowing of inferior vena cava could be detected in 40% of cases. Examination of the terminal parts of the hepatic veins demonstrated their occlusion or attenuation, a finding which is considered a new and significant contribution to the early diagnosis of this syndrome [14].

4.3. Inferior vena cava angiogram:

Presented as narrow or closed intra-hepatic portion of the inferior vena cava with marked collaterals [14].

4.4. Liver biopsy:

In the acute stage it shows centrilobular hemorrhage, necrosis and sinusoidal dilatation. In the chronic stage it presents picture of micronodular cirrhosis with normal portal tracts [7].

4.5. Other tools of investigations [14]

- The ascitic fluid is a main laboratory field of investigations. It usually shows protein values ranging between 1-3.5 gms/dl with occasional lymphocytes.

- Other sophisticated modules of investigations might be carried out : liver isotopic scanning, splenoportal venogram and arterio-venography of the portal system.

5. Management of Hepatic veno-occlusive disease:

No effective therapy until now especially in this type of Egyptian children. The target of available line is, may be, to reduce the complications, to reduce the stress of the patients and keep the patients in nearly comfortable life, but the following measures could be used safely [14].

5.1. Preventive measures:

- More investigation for the etiology of the disease especially pyrrolizidine alkaloids.

- Encouraging the breast feeding for two years as Glorious Qura'n says. (Sorra El bakara), regulation and careful inspection of diet after weaning [11].

- Good nutrition of the mother

- What about copper utensils ?? it suspected to play a role in indian cirrhosis !

5.2. Conservative measures :

- Follow up, because a grossly abnormal scan of liver and spleen in a patient with HVOD has been normalized completely without any interference.

- Colonic lavage to wash out the toxic metabolites.

5.3. Medical treatment:

- Low doses of *heparin or anticoagulants*, adapted dose of *prostacyclin.*

- *Anti inflammatory* or *steroids.*

- Use of *Vit C, u*se of *Vit. E* and Glutamine (source of glutathione) as antioxidants [15].

- Use of *recombinant tissue plasminogen activator* (rtPA), especially in patients after BM transplantation, *Urokinase* especially in cases with bleeding diathesis leading to thrombotic HVOD [15], [16], [17].

- *Diuretics* for ascites.

- *Large doses of glucose* together with *insulin* to aid glycogen deposition in the liver and so help its nutrition.

- Cupper chelation treatment (Di-penecillamine)

5.4. Surgical treatment [18].

5.4.1. Treatment of ascites :

- Frequent aspiration (partial or full)

- TIPS (transjagular intrahepatic portosystemic shunt)

- Hepatic and portal decompression for interactable ascites.

- LeVeen, peritoneojagular shunt.

5.4.2. Treatment of portal hypertension:

- Porto-systemic shunt as porto-caval, spleno-renal or meso-atrial.

- Acute venous obstruction could be treated by hepatofugal portal flow via veno-venous bypass to drain arterial blood flow.

5.4.3. Liver transplantation:

- It is now a part of the therapeutic armamentarium for this condition.

6. Therapeutic paracentesis [21].

The first study re-evaluating paracentesis as a treatment of cirrhotic patients with ascites consisted of a randomized controlled trial comparing repeated large-volume paracentesis (4-6 l/day until the disappearance of ascites) plus intravenous albumin infusion (40g after each tap) with standard diuretic therapy (frusemide plus spironolactone) in I17 patients with tense ascites and avid sodium retention who were admitted to several hospitals in the Barcelona area. This study, later confirmed by two more trials performed in Milan and Barcelona, showed the following results:

1. paracentesis was more effective than diuretics in eliminating ascites (96.5 versus 72.8%);

2. paracentesis plus albumin infusion did not induce significant changes in hepatic and renal function, serum electrolytes, cardiac output, plasma volume, plasma renin activity and plasma concentration of noradrenaline and antidiuretic hormone.

3. the incidence of hyponatremia, hepatic encephalopathy and renal impairment was much lower in patients treated with paracentesis.

4. the duration of hospital stay was lower in patients treated with paracentesis.

5. there were no significant probability of re-admission, probability of survival and causes of death between the two groups of patients.

Tito et al., later investigated whether ascites can be safely mobilized by total paracentesis (complete removal of ascites by a single paracentesis) plus intravenous albumin infusion

(6-8 g/l removed) in a one day hospitalization regime. The incidence of complications and the clinical course of the disease, as estimated by the probability of readmission to hospital, causes of re-admission, probability of survival and causes of death, were comparable to those reported by the same group of investigators in patients treated with repeated large-volume paracentesis.

In conclusion, these studies demonstrate that mobilization of ascites by paracentesis associated with intravenous albumin infusion does not impair systemic haemodynamics and renal function in patients with cirrhosis and tense ascites. Therapeutic paracentesis should be the treatment of choice for cirrhotic patients admitted to hospital with tense ascites, because it is more effective in mobilizing associated with a lower incidence of complications and reduce the duration of hospitalization. To avoid re-accumulation of ascites, patients treated with paracentesis require dietary sodium restriction and administration of diuretics after the procedures.

Subsequently, a trial was performed to establish whether intravenous albumin infusion is necessary in cirrhotic patients with tense ascites treated with repeated large-volume paracentesis. It was observed that paracentesis plus intravenous albumin does not induce significant changes in standard renal function testes, plasma renin activity and plasma aldosterone concentration. In contrast, paracentesis without albumin was associated with a significant increase in blood urea nitrogen, a marked elevation in plasma renin activity and plasma aldosterone concentration, and a significant reduction in serum sodium concentration. The number of patients developing hyponatremia and renal impairment was remarkably higher in patients treated with repeated large-volume paracentesis without intravenous albumin infusion. There are two detailed investigations assessing the effects of large-volume paracentesis without albumin infusion on systemic haemodynamics vasoactive hormones and renal function. A significant increase in cardiac output was observed 1 hour after treatment in both studies. Some hours later, however, a significant drop below baseline values was observed in cardiac output, pulmonary wedge capillary pressure and central venous pressure. Plasma renin activity increased and plasma atrial natriuretic peptide concentration decreased. The adverse effects observed after complete mobilization of ascites by paracentesis without albumin expansion did not occur in patients in whom ascites was only partially mobilized by paracentesis without colloid replacement. In conclusion, these studies demonstrate that complete mobilization of ascites by paracentesis without plasma volume expansion is followed by a reduction in effective intravascular volume, which leads to activation of the renin-aldosterone system and may impair renal function. The infusion of intravenous albumin is an important measure to prevent these abnormalities in cirrhotic patients with tense ascites treated with large-volume or total paracentesis.

Five randomized controlled trials and one prospective study aimed at investigating whether albumin can be substituted by less expensive plasma expanders (dextran-70, dextran-40, Haemaccel 5% and isotonic saline) have recently been reported. It has been observed that total or repeated large-volume paracentesis associated with intravenous administration of dextran-70 or Haemaccel is not associated with significant changes in renal and hepatic function. The incidence of hyponatremia, renal impairment and hepatic encephalopathy in patients receiving dextran-70 or Haemaccel was comparable with that in patients receiving albumin.

In one study, patients treated with dextran-70 showed a significant increase in plasma renin activity and aldosterone concentration. In a more recent study, however, therapeutic paracentesis plus intravenous dextran-70 administration was not associated with significant changes in plasma renin activity, which was measured 24 and 96 hours after the treatment. Cabrera et al., in one study including 14 patients, have suggested that intravenous isotonic saline infusion can also be a safe and cost effective alternative plasma expander in cirrhotics with tense ascites treated with paracentesis. Further studies are obviously needed to confirm their findings. It seems that dextran-40 is not as effective as albumin in preventing renal and electrolyte complications after therapeutic paracentesis, as renal impairment and/or hyponatremia developed after treatment in a relatively high proportion of patients.

Recently, a multicenter randomized trial comparing therapeutic paracentesis with PVS in cirrhotic patients with refractory or recurrent ascites has been published. More than 40 patients were included in each group. Both treatments were equally effective in mobilizing the ascites during the first hospital stay, although the duration of hospitalization was significantly longer in the shunt group. There were also no significant differences between both groups in the number of patients who developed complications or died. The number of re-admissions for any reason or for ascites, was significantly higher, and the time to first re-admission for any reason and for ascites significantly shorter in the paracentesis group than in the shunt group. The total time in hospital during follow-up, however, was similar in the two groups. The probability of shunt obstruction was 40 % at 1 -year follow-up. The probability of survival was similar in both groups. In conclusion, this trial shows that, although the LeVeen shunt was better than paracentesis in the long-term control of ascites, it did not reduce the total time in hospital nor prolong survival. On the other hand, patients treated with PVS required frequent re-operations due to obstruction of the prosthesis. Therapeutic paracentesis is therefore an alternative treatment to LeVeen shunt in cirrhotic patients with refractory ascites.

7. Peritoneovenous Shunting

In 1974 LeVeen [19], and colleagues developed a pressure-activated one-way valve for use in a peritoneovenous shunt (PVS). This device consists of a perforated intra-abdominal tube connected through a one-way pressure sensitive valve to a silicone tube that traverses the subcutaneous tissue up to the neck, where it enters one of the jugular veins (usually the internal jugular vein). The tip of the intravenous tube is located in the superior vena cava, near the right atrium or in the right atrium itself. The shunt produces a sustained circulating blood volume expansion by continuous passage of ascitic fluid to the general circulation. Flow in the shunt is maintained if there is a 3-5 cm H2O pressure gradient between the abdominal cavity and the superior vena cava. A loss of this gradient causes the valve to close, preventing blood from flowing back into the tubing. Two additional shunts have been introduced Denver and Cordis-Hakim. These latter shunts include a pumping mechanism that allows flow to be increased or a partially occluded shunt to be cleared.

The intravenous infusion of ascitic fluid through the shunt is associated with an increase in circulating blood volume and cardiac output. Since arterial pressure does not rise, there is a concomitant reduction in peripheral vascular resistance. These hemodynamic changes are associated with an increase in the plasma concentration of atrial natriuretic factor and a suppression of plasma levels of renin, aldosterone, noradrenaline and antidiuretic hormone. Urine volume and free water clearance increase in most patients. However, there is significant natriuresis in less than half of the patients, demonstrating that the PVS does not completely correct the abnormal sodium-retaining state associated with cirrhosis. Finally, in cirrhotic patients with moderate FRF, the PVS may improve renal blood flow and glomerular filtration rate. These hemodynamic and hormonal changes persist in most cases and a significant proportion of patients remains with minimal or no ascites despite a moderate sodium restriction and low diuretic dosage. There are also two studies that suggest that PVS has a positive effect on the nutritional status of patients in whom the shunt functions for a prolonged period of time. Despite these positive effects of PVS, there are a large number of complications, which may occur early in the postoperative period or at any time during follow-up [19], [20].

The role of PVS in the management of cirrhotic patients with ascitcs is still not well established. Only one prospective study showed that PVS is superior to conventional medical therapy in the management of ascites and in improving survival. By contrast, four randomized studies have failed to demonstrate a longer survival time in cirrhotic patients with ascites treated with PVS compared with medical therapy. Of these studies, that which was performed by Stanley et al., 1989 [22], is worth mentioning. They compared PVS with medical treatment (diuretics and occasional paracentesis) in 299 patients with cirrhosis and refractory or recurrent ascites. Although early mortality and probability of survival after randomization were similar in both therapeutic groups, PVS was more effective in the management of ascites than was conventional medical therapy, as indicated by shorter duration of first hospitalization, longer time to recurrence of ascites, and lower diuretic requirements during follow-up. However, these results are not surprising, because PVS was compared with a treatment that by definition was known to be ineffective.

The effect of PVS on survival in patients with FRF has also been studied in a randomized controlled trial. The treated patients had some improvement in renal function, but their survival was unaffected. Several studies have shown that morbidity and survival of cirrhotic patients treated with PVS correlate with the degree of impairment of liver and renal function. Therefore, the best results with this procedure should be expected to occur in those few patients with diuretic-resistant ascites and preserved hepatic function [23].

7.1. Early complications of peritoneovenous shunting

Acute bacterial infection is the most serious early complication. Staphylococcus aureus is a frequent isolate and represents the operative contamination of the shunt in some cases. The prosthesis is usually colonized and the infection cannot be eradicated in most cases unless the shunt is removed a high mortality can be expected. The prophylactic administration of anti-staphylococcal antibiotics 24 hours before and 48 hours after surgery reduces the inci-

dence of early postoperative infection. Biochemical disseminated intravascular coagulation (DIC) is seen in practically every cirrhotic patient treated with PVS in the early postoperative period. Bleeding caused by DIC develops most commonly in those patients with severe liver disease, but is now very uncommon, because many surgeons remove the ascitic fluid before inserting the shunt and replace it with normal saline. DIC is thought to develop because of infusion of factors present in ascitic fluid that activate coagulation (thromboplastin, activated clotting factors, endotoxin, collagen, plasminogen activator and fibrin split products). Postoperative fever, probably related to the passage of endotoxin contained in the ascitic fluid to the general circulation, is almost a constant and disappears spontaneously within the second postoperative week. Rapid expansion of the plasma volume is associated with a rise in portal pressure and may increase the risk of variceal haemorrhage. This complication can also be prevented by removing most ascitic fluid before the insertion of the shunt [24].

7.2. Long-term complications of peritoneovenous shunting

Obstruction of the shunt is the most common complication during follow-up. It occurs in more than 30% of patients and is usually due to deposition of fibrin within the valve or the intravenous catheter, thrombotic obstruction of the venous limb of the prosthesis, or thrombosis of the superior vena cava or right atrium initiated at the venous end of the shunt or damaged endothelium. Shunt obstruction is generally associated with ascites re-accumulation. Shunt patency can be assessed by Doppler ultrasound or by technetium 99m scintigraphy using intraperitoneal radioisotope injection. If the obstruction is confirmed, a shuntogram after the injection of contrast into the proximal limb of the shunt may identify the site of obstruction. Venography or digital angiography is necessary in the case of obstruction of the venous tip of the shunt. Superior vena cava syndrome secondary to total obstruction of the vein and pulmonary embolism are much less common. It is not clear that the insertion of a titanium tip into the venous end of the LeVeen shunt prevents thrombotic obstruction and the development of superior vena cava thrombosis. Finally, another long-term complication of PVS is small-bowel obstruction, which occurs in approximately 10% of patients and is due to intraperitoneal fibrosis [25].

8. Transjugular intrahepatic portosystemic shunt (TIPS)

The feasibility of intrahepatic portosystemic shunting was first demonstrated by Rosch and colleagues 1969 in pigs. Colapinto et al; 1982 [27] reported the first application of this technique to humans. This was attempted following transhepatic obliteration of varices in 20 severely ill patients with variceal hemorrhage. The authors inflated a balloon catheter in the intrahepatic track and left it there for 12 hours. In an initial report all six shunts studied were patent 12 hours after the procedure and one was still patent at autopsy 6 weeks late.

Many demonstrated prolonged patency of the shunt for up to 10 months and ease of recanalizing the radiopaque shunt when occlusion occurred. This expandable stent was then used

successfully in patients with portal hypertension. Similar good results were soon reported with the self-expanding Wall stent. Percutaneous portography was used in the early cases to facilitate transjugular portal vein puncture. With increasing experience this has been replaced by ultrasound guidance in most centers [28].

There is now an increasing array of equipment available for transjugular intrahepatic portosytemic shunt (TIPS) insertion. The most widely used needles are a standard transjugular biopsy needle with a straight or reversed bevel (Cook Ltd) or the Richter needle which has a tapered tip and a blunt obturator (Angiomed, Karlsruhe, Germany). Another set with a blunt cannula, through which is passed a sharp style is also available (Cook). There is also a wider choice with regard to the type and dimensions of metal stent. In addition to the original Palmaz and Wall stents, there is the Strecker stent and the Memotherm stent (Angiomed, Karlsruhe, Germany). Claimed advantages for these new stents are increased radioopacity (Strecker stent) and improved delivery systems (Memo stent) [29].

A recent randomized controlled study compared the Palmaz and Wall stent in 90 patients and found little difference in outcome. Early shunt thrombosis was more likely with the Wall stent (9%), whereas stenosis of the hepatic vein was more likely with the Palmaz stent (I3%). Experience with the other stents is limited.

As yet the long-term expectations of TIPS have not been fulfilled in those clinical situations in which long-term efficacy is needed as prevention of variceal rebleeding, ascites, cirrhotic hydrothorax, Budd-Chiari syndrome, and long-term amelioration of clinical status before liver transplantation. All these indications need controlled trials against current best optimal management before TIPS is used routinely even for an individual patient. The high stent obstruction rate is the most important limiting factor, but change in stent shape, coating material or other technical aspects may overcome this [30].

The complications of TIPS are significant if elective and long-term use is considered, thus the need for trials before new therapies are introduced. In an emergency situation the complications due to TIPS are an acceptable risk, but again information from controlled trials is needed. This is particularly true when TIPS is used as a short-term bridge to liver transplantation. TIPS will have a place in the treatment of cirrhotic patients. At present short-term rather than long-term indications appear to be where TIPS will have more beneficial effects [28].

9. Liver transplantation: and hepatic venous obstruction

Liver transplantation for Budd–Chiari syndrome: A European study on 248 patients from 51 centers) [31]: The results of liver transplantation for Budd–Chiari syndrome (BCS) are poorly known and the role and timing of the procedure are still controversial. The aim of this study was to investigate the results of transplantation for BCS, focusing on overall outcome, on prognostic factors and on the impact of the underlying disease. Methods: An enquiry on 248 patients representing 84% of the patients transplanted for BCS in the European Liver Transplantation Registry between 1988 and 1999. Results: Of the 248 patients, 70.4% were

female and 29.6% male. The mean age was 35.7 years. The overall actuarial survival was 76% at 1 year, 71% at 5 years and 68% at 10 years. 77% of deaths occurred in the first 3 months: 47% were due to infection and multiple organ failure, and 18% to graft failure or hepatic artery thrombosis. Late mortality (>1 year) occurred in nine patients, due to BCS recurrence in four of them. The only pre-transplant predictors of mortality on multivariate analysis (Cox) were impaired renal function and a history of a shunt.

10. Conclusions

Liver transplantation for BCS is an effective treatment, irrespective of the underlying cause, and should be considered before renal failure occurs [31].

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