Available online at www.scientiaresearchlibrary.com

Scientia Research Library

ISSN 2348-0416
USA CODEN: JASRHB

Journal of Applied Science And Research, 2018, 6 (6):18-26

(*http://www.scientiaresearchlibrary.com/arhcive.php*)

Plasmapheresis in severe form of HELLP Syndrome : interesting therapeutic perspective

-clinical case and review of the literature-

Adil ZEMMOURI(1) , Abdelghafour EL KOUNDI(1) , Hicham EL FAZAZI(2), Mohamed DEHAYNI(2) , Lahcen SAFI(1)

(1) Intensive care unit ,Mohammed V Military hospital , Rabat ,Morocco

(2) Gynecology unit ,Mohammed V Military hospital , Rabat ,Morocco

---

ABSTRACT

HELLP Syndrome is a pathology of the pregnant woman. It is defined by biological parameters associating the triad: Acute haemolysis with anemia, thrombocytopenia and hepatocytolysis. HELLP syndrome is a rare condition (0.6% of pregnancies) [1]... It can be serious because of acute renal failure and the bleeding disorders it can cause. Maternal mortality is 2-40% and neonatal mortality is 7-60%[2]. Serious forms require multidisciplinary care. The purpose of this observation is to report the different current techniques that improve the prognosis of HELLP Syndrome in its severe form.

INTRODUCTION

Interest

Through this work we try to highlight through 3 clinical observations some aspects that could take the OHSS in its critical form. This syndrome often unknown in its mild and moderate forms is in its severe form a threat that can be life-threatening, it is a complication that we may see more and more in our practice because of the explosion of practice IVF (currently 19 centers)

MATERIAL AND METHODS

Observation :

This is a patient aged 36, admitted for the management of preeclampsia with biological complications at 33 weeks of amenorrhea.

The patient presented a completed pregnancy 10 years ago with uncomplicated vaginal delivery. The current pregnancy was badly followed in a private health facility. During the interrogation, no pathological antecedent is found, in particular no renal, hepatic or autoimmune pathology (Systemic Lupus Ewthematosus, Autoimmune Thrombocytopenic Purpura). The patient presented to gynecological emergencies on the night of July 25, 2006 for epigastric bar pain, headache and myodopsia. At the clinical examination, we find an arterial hypertension at 195/110 mmHg, edema of the lower limbs going back to mid-thigh and the urine test strip showed proteinuria. The

diagnosis of severe preeclampsia was therefore made.

Obstetrical ultrasound examines a retroplacental hematoma. Emergency fetal extraction by caesarean section was therefore performed under general anesthesia. The newborn weighed 1250g at 33 weeks of amenorrhea. Just before the gesture, the patient had a seizure that was treated with VALIUM 5mg direct intravascular.

Subsequently, the patient was transferred to the intensive care unit for coagulation disorders with 40% PT, 12.900 / ml deep thrombocytopenia and the search for PDF was positive. She also had biological renal failure with urea at 0.54 mg / l and serum creatinine at 28 mg / l. The patient was transfused with 10 pellets and 8 units of fresh frozen plasma to curb his bleeding disorders

At admission, the patient was conscious, eupnetic. The blood pressure was 130/100 mm Hg under LOXEN at the electric syringe push. The rest of the exam was peculiar.
The immediate course was marked by two episodes of concomitant tonic-clonic convulsive seizures with two hypertensive peaks (210/120 and 160/100) controlled by 5 mg of VALIUM in IVD. Biologically, we found:

-Hemolytic anemia with 6.8g / dl hemoglobin and blood smear schistocytes with LDH 0 598 IU / l.

-Thrombocytopenia at 12,600 / ml.

-Hepatic cytolysis with ASAT 0 2064 IU / l and ALAT at 608 IU / l.

The diagnosis of severe form of HELLP syndrome was retained.

The patient was treated as follows:

-MAGNESIUM SULFATE: 10 ampoules daily for 2 days to prevent seizures.

-DEXAMETHASONE: 6mg / 6hours with a gradual decrease over 9 days to block the activating factors.

-LOXEN to push the electric syringe with a dosage adapted according to the blood pressure to stabilize the blood pressure.

On day 1: worsening of abdominal pain and ascites on clinical examination were noted. Abdominal CT without injection was performed and showed a large peritoneal effusion of all cavities as well as bilateral pleural effusion. The ascites puncture brought back 2L of serumematic fluid

At D2: deterioration of respiratory and renal function. Appearance of acute clinical and radiological and gasometric lung edema which required tracheal intubation and artificial ventilation. The patient was sedated by combination of HYPNOVEL and FENTANYL at low dose.

On the renal plane, the patient is oligo-auric (dark urine with hemoglobin deposits). Due to aggravation of renal function (Urea at 2g / l and creatinemia at 80 mg / l). it was decided to perform daily hemodialysis sessions for 6 days associated with transfusions of 8 packed red cells. 16 fresh frozen plasmas and 20 platelet concentrates.

Two plasmapheresis sessions were performed every other day to purge the etiopathogenic factors of the HELLP syndrome, according to the data of the literature

At D10: clinical improvement was observed with resumption of diuresis, improvement of clinical respiratory, radiological and gasometric status. This allowed us to wean the patient from artificial ventilation and ex tuber 12 hours later. Moreover, the biological parameters have gradually

normalized (see curves).

The patient was transferred to the gynecology department after 13 days in intensive care. The treatment consists of an oral antihypertensive agent, a low molecular weight heparin and a diuretic.

The patient was seen at day 35, she is doing well outside a persistent hypertension at 160/90 mmHg balanced by LOXEN LP.

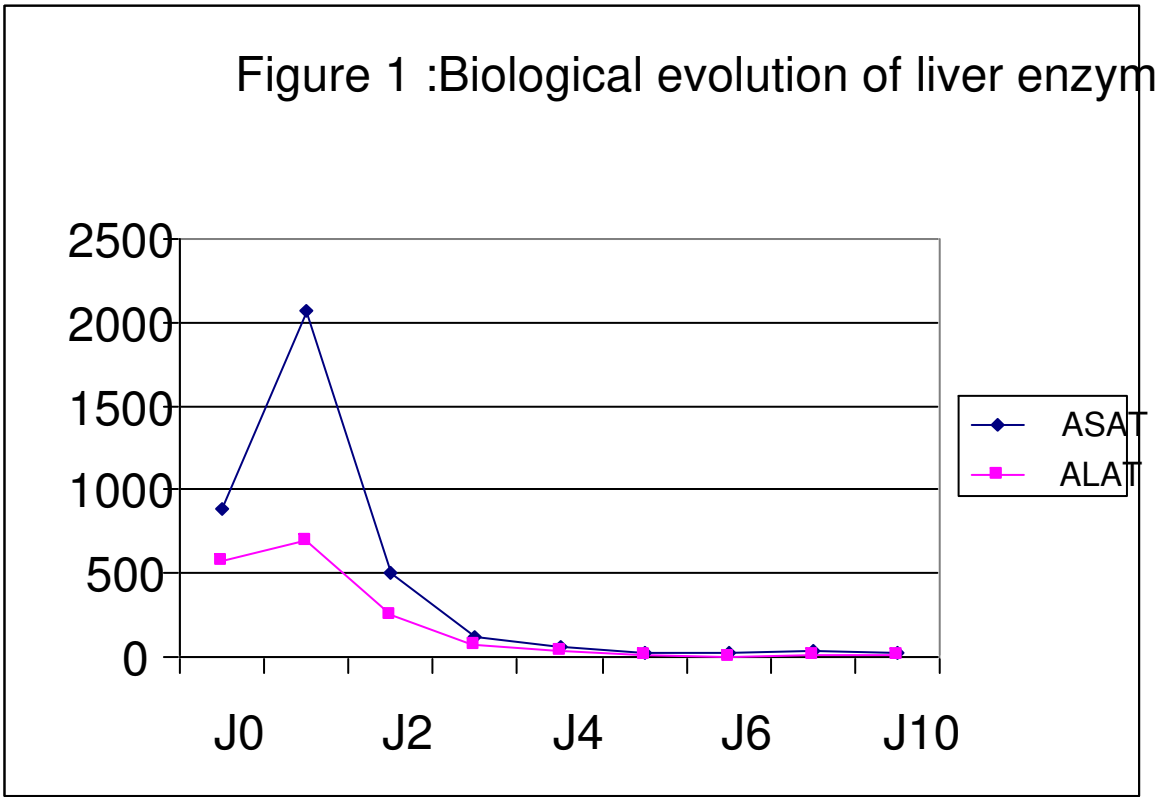


Figure 1 :Biological evolution of liver enzymes
2500
2000
1500
1000
500
0
J0
J2
J4
J6
J10
ASAT
ALAT

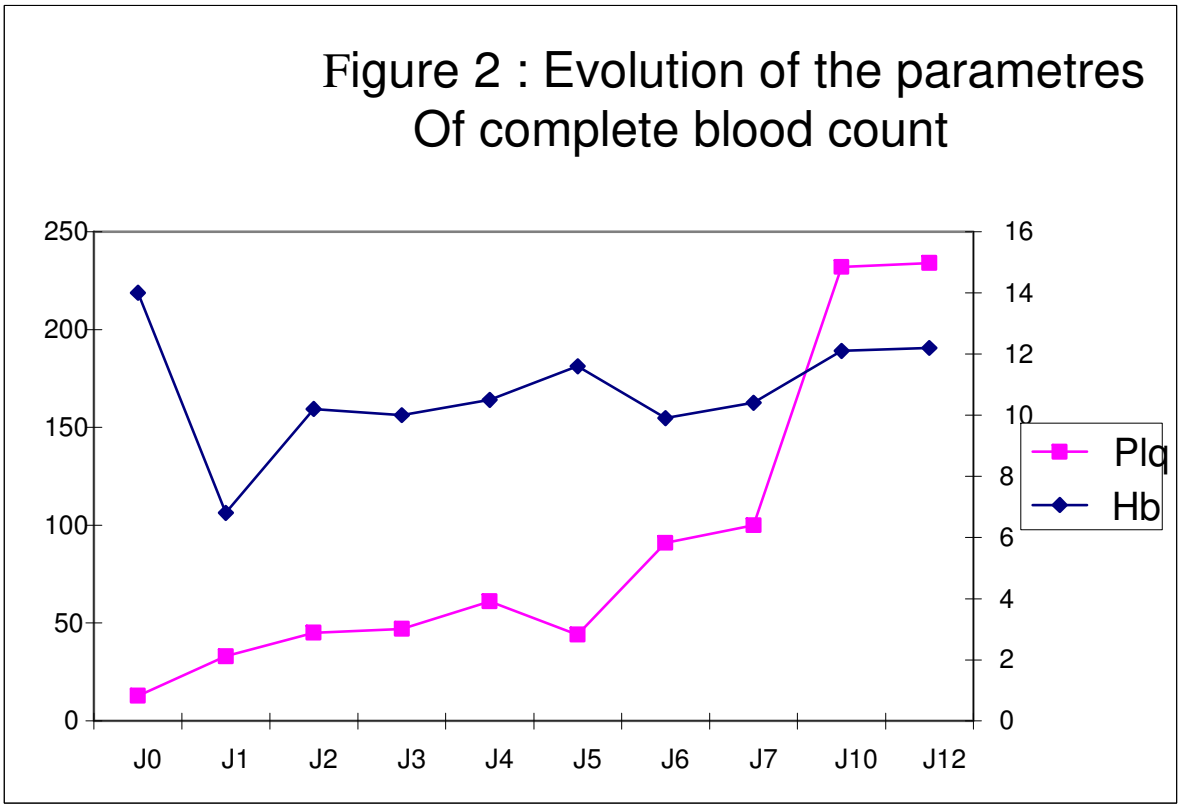
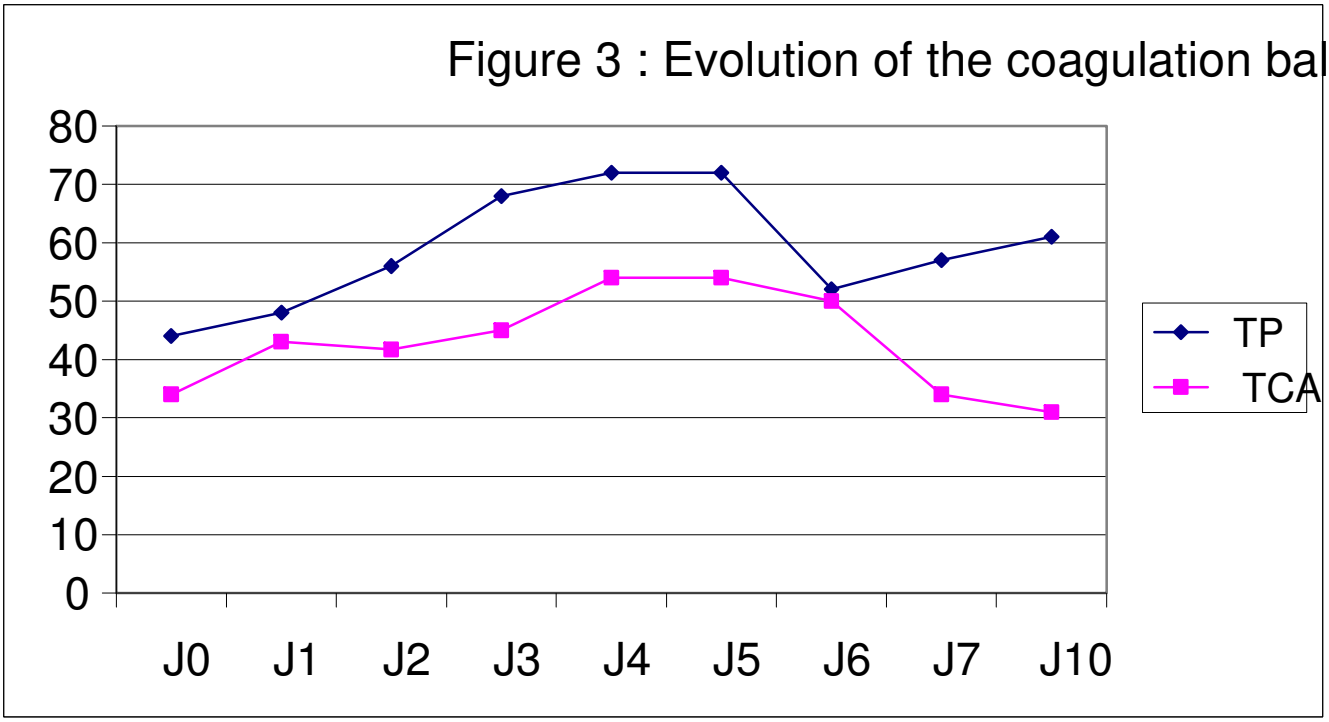


Figure 2 : Evolution of the parametres
Of complete blood count
250
200
150
100
50
0
16
14
12
10
8
6
4
2
0
J0
J1
J2
J3
J4
J5
J6
J7
J10
J12
Plq
Hb



RESULTS AND DISCUSSION

This syndrome was described for the first time, quite late in 1982 by weinste in[3]. This term is an acronym that associates hemolysis (Hemolysis), hepatic cytolysis (Elevated liver enzyme) and thrombocytopenia (Low platelets). We therefore note that this syndrome has only a biological definition.
The definition of the syndrome will remain unclear for many years until a stricter definition by Sibai in 1990[4]. It differentiated HELLP complete syndrome, partial HELLP syndrome and eclampsia mellitus without biological abnormality.

The current definition is still purely biological and relies on the triad which, if completely present, defines the complete HELLP Syndrome. If one or two elements are missing, the HELLP Partial or Incomplete Syndrome is defined. If no biological abnormality is found in a context of severe pre-eclampsia, it is called severe pre-eclampsia without biological abnormality.

We must therefore find:

- Acute mechanical hemolysis responsible for peripheral anemia often poorly tolerated because of rapid installation.

-Peripheral thrombocytopenia

- Hepatic cytolysis with elevated transaminases (which may also be related to acute hemolysis)

This triad is most often found in a context of pre-eclampsia.

This pathology is rare, it complicates only 0.6% of pregnancy[1-2], 50% of cases are reported between 27 and 36 weeks of amenorrhea. 20% of cases are early between 17 and 20 weeks of amenorrhea and 30% of cases can occur during the postpartum between the 1st and 4th day[2].

Clinical signs found in pre-eclampsia[5]:

- An epigastric pain in the so-called DE bar slipper in 65 to 90% of cases.

-Digestive signs such as nausea, vomiting, diarrhea.

-A high blood pressure.

-Signs related to high blood pressure with neurosensory signs (headaches, tinnitus, visual disturbances ...)

-Edema of the lower limbs.

It should be noted that in 15% of the cases there is no sign before runner not including arterial hypertension[2].
We can find the signs related to acute hemolysis: jaundice, hemorrhagic shock, dark urine ..., signs of complications.

We find in the HELLP Complete Syndrome:

-Acute anemia related to mechanical hemolysis with lower hemoglobin, onset of LDH, free bilirubin.

-Peripheral thrombocytopenia less than 100.000 / ml.

-Hepatic cytolysis with an increase of transaminases above 70 IU / ml[4].

Associated with these biological signs, we find the signs related to preeclampsia with urinary strip proteinuria, an increase in uric acid and the signs related to x complications (renal failure with increased urea and creatinine, disseminated intravascular coagulation with disruption of coagulation blian ...)

Three forms of this syndrome are described according to the presence or not of all the biological criteria of the triad[4].

If all the criteria are present, we have the complete form of the HELLP Syndrome. This is considered severe if the thrombocytopenia is less than 30000 / ml or if the transaminases are greater than 5000 IU / ml.

If one or two criteria are missing, it is called severe pre-eclampsia without biological abnormality. This distinction is necessary because the prognosis is different as well as the care.

About maternal complications according to an article by François Audibert et al. Performed between 1992 and 1995 out of 316 women admitted for severe pre-eclampsia, 21% (67 women) were found with HELLP Complete Syndrome, 22% (71 women) with HELLP Partial Syndrome and 56% (176 women) with severe plasmapheres is without abnormality biological[5].

The subscapular hematoma of the liver is a serious complication that deserves to be systematically sought by an abdominal ultrasound because its rupture causes a fulminating haemorrhagic shock and the risk of mortality is important. This hematoma is most often located in the superior anterior part of the right hepatic lobe. The risk factors for this complication are high age and multiparity.

About neonatal complications [7] they are related to prematurity and therefore depends on the age and weight of the infant. Indeed, the more the child is born prematurely the greater the risks. There are complications that are not specific to children born to mothers with HELLP Syndrome. We find the hyaline membrane disease, intraventricular hemorrhage, necrotizing ulcerative enterocolitis and of course iatrogenic complications and infant death (in 7 to 60%).

If the pregnancy has passed 32 weeks of amenorrhea, the termination of pregnancy is recommended by triggering vaginal delivery unless there is a vital risk for the mother (DIC, uncontrolled hypertension, eclampsia ...) or for the fetus (acute fetal distress, retro placental hematoma, etc.), to which delivery will be by caesarean section.

Before 28 weeks of amenorrhea or if there is no risk for the woman and the clinico-biological parameters improve, we can move towards a conservative management with strict supervision of the mother and the fetus in a level III maternity ward.

Between these dates it is necessary to transfer the patient to a maternity level III by administering corticosteroid therapy (DEXAMETHAZONE), it will allow lung maturation and clinical-biological improvement of the mother[1-11].

The mother and her child will then be cared for in a resuscitation department with experience in this area.

In intensive care, it will be necessary:

-Control blood pressure with NICARDIPINE alone or in combination with CLONIDINE.

-Control convulsions with DIAZEPAM and prevent recurrence with MAGNESIUM SULFATE (which also reduces morbidity) at the dose of 6g in loading dose then 2g / hour for 48 hours[1].

-If necessary to control ventilation by intubation and artificial ventilation.

-Assure a good filling according to the central venous pressure and the contribution of anemia. We must take care of the various complications:

-Acute kidney failure oligo-auric: this is only transient and requires only transient hemodialysis. No

cases of chronic insufficiency have been described[8].

Intra-vascular Disseminated Coagulation : Transfusion of fresh frozen plasma and platelet concentrate can be performed during hemodialysis[9].

New perspectives are currently being studied:

Plasma exchanges: they must be realized after the delivery and more firstly possible. This therapy allows a reduction of the mortality, the duration of stay in intensive care and the normalization of the different biological parameters previously disturbed. This technique would rather be reserved for HELLP Refractory Syndrome or having one or more organ failures[10].

- Corticosteroid therapy with DEXAMETHAZONE is the subject of much research. She would have high doses (10mg every 12 hours) of benefits for the clinical symptoms (mainly on epigastric pain and oligo-anuria) and the biological parameters of the mother. But it would have no benefit on morbidity and mortality (retro placental hematoma, pulmonary edema and subcapsular hematoma of the liver) and the need for transfusion. It allows fetal lung maturation but provides no benefit over normal doses of corticosteroids on fetal complications. It would delay the onset of labor, thus reducing prematurity[11]. This perspective still needs to be studied

The multiplicity of possibilities requires cooperation between the various services involved and all the intensive care unit staff who must be trained and have experience in the field to improve the care of patients.

This syndrome is rare but of great severity for the patient and her child.

For the mother[12]: the mortality varies between 2 and 40%. This mortality is all the more important that the diagnosis is made late, that the syndrome appeared during pregnancy and also depends on the number of complications that the patient presents at admission. Obviously, this risk is reduced if the patient is cared for in an intensive care unit with experience in this area. If the delivery is done quickly, the risks decrease with a return to normal which varies between 7 and 14 days.

For the fetus[13]: perinatal mortality is important from 7 to 60%. This occurs most often by complications related to prematurity. It depends on the management of the mother in ante-partum, the date of prematurity and the experience of the service in the care of infants born to mothers with this syndrome.

CONCLUSION

HELLP syndrome is a severe gravid-puerperal complication that requires immediate termination of pregnancy if there are serious maternal complications. Speed of diagnosis, emergency fetal extraction and medico-surgical management allow rapid clinical and biological improvement. The range of possibilities is widening with the arrival of new therapeutic means that still need to be studied to determine their place in the therapeutic strategy.

REFERENCES

[1] M.D. Bèye, E.Diouf, M.D. Bah, M. Ndoye-diop, O. Kane and B.Sall-ka, Prise en charge du HELLP Syndrome en réanimation à Dakar, Annales d'anesthésie et de réanimation Vol.25, Issue 3, Mars **2006**, pp 291-295.

[2] Tsarsais.V, Audibert. F, le HELLP Syndrome

[3] Weinstein L. Syndrome of hemolysis, elevated liver enzymes and low platelet count: a severe consequence of hypertension in pregnancy. AmJ Obstet Gynecol **1982**;142(2):159Ŕ67.

[4] Sibai BM. The HELLP syndrome (hemolysis, elevated liver enzymes and low platelets): much ado about nothing? Am J Obstet Gynecol **1990**;162 (2):311

[5] Audibert F, Friedman SA, Frangieh AY, Sibai BM. Clinical utility of strict diagnostic criteria for the HELLP syndrome. Am J Obstet Gynecol **1996**;175:460-4

[6] Ana CPF, Araujo MD, Ph D, Marcos D. Et al, Characteristics and treatement of hepatic rupture cused by HELLP syndrome Am J obstet Gynecol **1996** pp 460-464

[7] Dorel Abraovici MD, Steven A. Freidman MD et al, Brian M. Mercer MD, François Audibert MD, Lu Kao RN and Baha M. Sibai MD, Neonatal outcome in severe preeclampsia at 24 to 36 weeks gestation : Does the HELLP Syndrome matter ?Am J obstet Gynecol **1999** pp 221-225

[8] Drakeley AJ, Le roux PA, Anthony J, Penny J, Acute failure complicating severe preeclampsia requiring admission to an intensive care unit. Am J obstet Gynecol **2002** pp 253-256

[9] Ellart D, Guevart E, Forzy G, House de l'Aulnoit D, Nabi A, Delcroix M, Brabant G, HELLP syndrome. A propos of series of 9 cases without disseminated intravaculair coagulation-Revue française de gynécologie et d'obstétrique **1990** pp220-226

[10] Bulent Eser, MD, Muhammet Guven, MD, Ali Unal, MD, * Ramazan Coskun, MD, Fevzi Altuntas, MD,* Murat Sungur, MD, Ibrahim Serdar Serin, MD,: The Role of Plasma Exchange in HELLP Syndrome Clin Appl Thrombosis/Hemostasis 11(2):211Ŕ217, **2005**

[11] Nunes F, Campos AP, Avillez T, Rodriques RQ, Meirinho M, Corticosteroid therapy for patients with HELLP Syndrome- Am J obstet Gynecol **2004** pp 263-268

[12] Malvino E, Maternel morbidity and perinatal mortality in HELLP Syndrome. Multicentric studies in intensive care units in buenos Aires area, Medecina (B Aires) **2005**,65(1) 17-23

[13] Padden MO, HELLP Syndrome : recognition and perinatal management. Am Fam Physician **1990** sep 1 ; 60(3) : 829-839.