If the patient is <34 weeks gestation and maternal and fetal status are

1. Obstetric management

A prospective national study of acute fatty liver of pregnancy in the UK. Gut.

• Criteria for AFLP from case series and expert opinion were used in a

• AFLP has overlapping features with HELLP, but there is no well-

established definition of the condition that clearly differentiates it from HELLP.

Criteria for AFLP from case series and expert opinion were used in a

• AFLP has overlapping features with HELLP, but there is no well-

established definition of the condition that clearly differentiates it from HELLP.

Differentiating between severe preeclampsia, HELLP, AFLP, or evolving

HELLP syndrome may be difficult (see Table 6).

While the use of echocardiography has been described in paroxysmal nocturnal

第五届世界胎盘功能障碍（HELLP）综合征国际会议

Corticosteroids—may improve the platelet count and other laboratory

parameters more quickly, but have not been shown to improve long-term

fetal or maternal outcomes.

During pregnancy, Adriamycin, methotrexate, and vincristine

are avoided.

VIII. Thrombotic Thrombocytopenia Purpura (TTP)/Atypical Hemolytic Uremic Syndrome (aHUS)

During pregnancy, Adriamycin, methotrexate, and vincristine

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During pregnancy, Adriamycin, methotrexate, and vincristine

are avoided.
I. Introduction to Thrombocytopenia in Pregnancy

- Thrombocytopenia is second to anemia as the most common hematologic abnormality encountered during pregnancy.
- The prevalence of a platelet count < 100 x 10^9/L in the third trimester of pregnancy is 60-70%.
- A platelet count of < 100 x 10^9/L, the definition for thrombocytopenia, is observed in only 1% of pregnant women.

The hematologist’s role is to:
- determine the cause
- advise in the management of thrombocytopenia
- help estimate the risk to the mother and fetus

II. Causes of Thrombocytopenia in Pregnancy

The hematologist is usually consulted in one of three scenarios:
1. Pre-existing thrombocytopenia in pregnancy, primary thrombocytopenia (ITP)
2. Decreasing platelet count or newly discovered thrombocytopenia in pregnancy, which may or may not be related to pregnancy
3. Acute onset of thrombocytopenia in the setting of severe preeclampsia

Thrombocytopenia is second to anemia as the most common hematologic abnormality encountered during pregnancy. However, in 15-20% of cases of HELLP syndrome, no hypertension or proteinuria is present. 70% of cases occur in the late second or third trimester, the remainder occur postpartum.

Table 2. Basic Laboratory Evaluation of Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Recommended tests</th>
<th>Implying infection</th>
<th>Implying autoimmune disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Viral screening (HIV, HCV, HBV)</td>
<td>Anti-nuclear antibody (ANA)</td>
<td>Anti-thrombopoietin receptor agonists (C)</td>
</tr>
<tr>
<td>Bone marrow biopsy</td>
<td>H. pylori testing</td>
<td>Campath-1H</td>
</tr>
<tr>
<td>Anti-platelet antibody testing</td>
<td>Direct antiglobulin (Coombs) test</td>
<td>Rituximab</td>
</tr>
</tbody>
</table>
| Reticulocyte count | Quantitative immunoglobulins | Anti-D immunoglobulin | [C]

**Note:** Relapsed contraindicated

Table 3. Therapeutic Options for ITP in Pregnancy

<table>
<thead>
<tr>
<th>First line therapy</th>
<th>Second line therapy for ITP refractory to first line therapy</th>
<th>Third line therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral corticosteroids</td>
<td>Combined corticosteroids and IgG</td>
<td>Relatively contraindicated</td>
</tr>
<tr>
<td>Oral corticosteroids</td>
<td>Corticosteroids and IVIg</td>
<td>First line therapy</td>
</tr>
<tr>
<td>Oral corticosteroids</td>
<td>Plasmapheresis</td>
<td>Second line therapy for ITP refractory to first line therapy</td>
</tr>
</tbody>
</table>

**Note:** Anti-D immunoglobulin [C] Relatively contraindicated

V. Management of ITP at the Time of Delivery

- Women with no bleeding manifestations and platelet counts ≥ 30 x 10^9/L do not require any treatment until 36 weeks gestation (or sooner if delivery is indicated).
- If platelet counts are < 30 x 10^9/L, regional anesthesia is preferred to dexamethasone, which crosses the placenta more readily.
- If platelet counts are < 30 x 10^9/L or clinically relevant bleeding is present, platelet transfusion in conjunction with IVIg can be considered.
- For a woman whose platelet count is < 30 x 10^9/L but has not required therapy during pregnancy, oral prednisone (or prednisolone) can be started before 36 weeks gestation.

**Note:** Medications are adjusted to maintain a safe platelet count (see below).

**Note:** While "The American Society of Hematology 2011 Evidence-Based Practice Guideline for Immune Thrombocytopenia" recommends a starting dose of prednisone of 1mg/kg daily, there is no evidence that a higher starting dose is better than a lower dose, and other experts recommend a starting dose of 0.25 to 0.5 mg/kg daily.
- Medications are adjusted to maintain a safe platelet count (see below).


**Note:** Oral corticosteroids [D] Vinca alkaloids [D] Thrombopoietin receptor agonists [C] Dapsone [C] Splenic sequestration (liver enzymes, elevated liver enzymes, low platelets) or AFLP (acute fatty liver of pregnancy)

VII. Acute Onset of Thrombocytopenia in the Setting of Severe Preeclampsia

1. Preeclampsia, which affects 5-8% of pregnant women, is diagnosed when:
   - systolic BP ≥ 140 mmHg or diastolic BP ≥ 90 mmHg is present or was present 24 hour prior to pregnancy. This can be defined as urinary excretion ≥ 0.3 g protein/24-hour
   - in a woman with previous normal blood pressure

2. Eclampsia is new-onset grand mal seizures in a woman with preeclampsia.

3. Superimposed preeclampsia may develop in a woman with a history of chronic hypertension and is manifested by:
   - development of proteinuria ≥ 0.3 g/day after 20 weeks of gestation
   - the development of hypertension after 20 weeks of gestation, or
   - the development of the HELLP syndrome

4. Severe preeclampsia is diagnosed when one or a number of different critical criteria are met. Approximately 0.5-1.5% of all women develop a platelet count < 100 x 10^9/L at term, while 0.3-0.5% have severe preeclampsia.

**Note:** Oral corticosteroids [D] Vinca alkaloids [D] Thrombopoietin receptor agonists [C] Dapsone [C] Splenic sequestration (liver enzymes, elevated liver enzymes, low platelets) or AFLP (acute fatty liver of pregnancy)


B. HELLP Syndrome

HELP syndrome, which affects 0.6-8% of pregnant women, is a variant of preeclampsia where ≥ 15% of cases of HELLP syndrome, no hypertension or proteinuria is present. 70% of cases occur in the late second or third trimester, the remainder occur postpartum.
I. Introduction to Thrombocytopenia in Pregnancy

- Thrombocytopenia is second to anemia as the most common hematologic abnormality encountered in pregnancy.
- The prevalence of a platelet count < 100 x 10^9/L in the third trimester of pregnancy is 6 to 10%.
- A platelet count of < 100 x 10^9/L, the definition for thrombocytopenia, is observed in only 1% of pregnant women.

The hematologist’s role is to:
- determine the cause
- advise in the management of thrombocytopenia
- help estimate the risk to the mother and fetus

II. Causes of Thrombocytopenia in Pregnancy

The hematologist is usually consulted in one of three scenarios:
1. pre-existing thrombocytopenia in the patient
2. decreasing platelet count or newly discovered thrombocytopenia in pregnancy, which may or may not be related to pregnancy
3. acute onset of thrombocytopenia in the setting of serious products

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2. decreasing platelet count or newly discovered thrombocytopenia in pregnancy, which may or may not be related to pregnancy
3. acute onset of thrombocytopenia in the setting of serious products

The hematologist is usually consulted in one of three scenarios:
I. Introduction to Thrombocytopenia in Pregnancy

- Thrombocytopenia is second to anemia as the most common hematologic abnormality encountered in pregnancy.
- The prevalence of a platelet count <100 × 10^9/L in the third trimester of pregnancy is 1-2%.
- A platelet count of <100 × 10^9/L is the definition for thrombocytopenia in pregnancy, adopted by the International Working Group, is observed in only 1% of pregnant women.

The hematologist’s role is to:

- determine the cause
- advise in the management of thrombocytopenia
- help estimate the risk to the mother and fetus

II. Causes of Thrombocytopenia in Pregnancy

The hematologist is usually consulted in one of three scenarios:

1. pre-existing thrombocytopenia in the mother (e.g. ITP)
2. decreasing platelet count or newly discovered thrombocytopenia in pregnancy, which may or may not be related to pregnancy
3. acute onset of thrombocytopenia in the setting of severe products (the HELLP syndrome (hemolysis, elevated liver enzymes, low platelets)) or AFLP (acute fatty liver of pregnancy)

Table 1. Causes of and Relative Frequencies of Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-existing</td>
<td>Autoimmune disorders</td>
<td>5-10%</td>
</tr>
<tr>
<td>Disease-related</td>
<td>Thrombocytopenia associated with systemic disorders</td>
<td>2-5%</td>
</tr>
<tr>
<td>Infections</td>
<td>Severe infections</td>
<td>1-2%</td>
</tr>
</tbody>
</table>

Table 2. Basic Laboratory Evaluation of Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Test</th>
<th>Description</th>
<th>Normal Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Platelet count</td>
<td>Number of platelets per microliter of blood</td>
<td>&gt;100 × 10^9/L</td>
</tr>
<tr>
<td>Bone marrow aspiration</td>
<td>Biopsy of bone marrow for evaluation of bone marrow</td>
<td>Normal appearing bone marrow</td>
</tr>
</tbody>
</table>

Thrombocytopenia is defined as a platelet count of <100 × 10^9/L at any time during pregnancy.

III. Decreasing Platelet Count or Newly Discovered Thrombocytopenia in Pregnancy: Gestational Thrombocytopenia

- Accounts for ~70-80% of cases of thrombocytopenia in pregnancy and is typically characterized by a platelet count > 70 × 10^9/L
- Commonly occurs in the mid-second to third trimester.

- No confirmatory labs: diagnosis of exclusion.
- Mechanism unknown, but hemodilution and accelerated clearance are postulated.
- No management is required, but platelet count < 70 × 10^9/L warrants an investigation for an alternative etiology.
- Typically resolves within 6 weeks postpartum, but may recur with subsequent pregnancy, therefore, not recommended.
- Not associated with neonatal thrombocytopenia.

IV. ITP and Its Management in Pregnancy

- Women with no bleeding manifestations and platelet counts ≥ 30 × 10^9/L do not require any treatment until 30 weeks gestation (or sooner if delivery is necessary).
- Platelets counts are < 30 × 10^9/L or clinically relevant bleeding is present, or trophoblastic disease, ITP is a diagnosis of exclusion.

- The recommended starting dose of IVIG is 1 g/kg.
- For a woman who is not pregnant, platelet count < 80 × 10^9/L has not required therapy during pregnancy, oral prednisone (or prednisolone) can be started 10 days prior to anticipated delivery at a dose of 10-20 mg daily and titrated as necessary.
- Given the difficulty predicting severe thrombocytopenia in neonates and the very low risk of intravascular hemolysis (<1% of births) or mortality (<1%), the need for delivery should be determined by obstetric indication.
- Percutaneous umbilical blood sampling (PUBS) or fetal scalp blood sampling is not helpful in predicting neonatal thrombocytopenia, or in the presence of postpartum thrombocytopenia, thrombocytopenia is to be managed expectantly.

- The infant nule platelet count occurs 2-6 days after delivery and a spontaneous rise occurs by day 7.

- Women with ITP are at an increased risk of venous thromboembolism, and some form of postpartum thrombocytopenia is to be managed expectantly.

References:


V. Management of ITP at the Time of Delivery

- Current recommendations aim for a platelet count of ≥ 50 × 10^9/L prior to labor and delivery as the risk of cesarean delivery is present with every delivery.
- The minimum platelet count for the placement of regional anesthesia is undefined but local providers typically use a platelet count ≥ 50 × 10^9/L for anesthesiaagogues will place a regional anesthetic if the platelet count is ≥ 80 × 10^9/L.
- While platelet transfusion is effective in ITP if an adequate platelet count has not been achieved and delivery is emergent, platelet transfusion in conjunction with IVIG can be considered.
- For a woman who is not pregnant, platelet count < 80 × 10^9/L has not required therapy during pregnancy, oral prednisone (or prednisolone) can be started 10 days prior to anticipated delivery at a dose of 10-20 mg daily and titrated as necessary.
The hematologist’s role is to:
• determine the cause
• help estimate the risk to the mother and fetus

### II. Causes of Thrombocytopenia in Pregnancy

The hematologist is usually consulted in one of three scenarios:
1. pre-existing thrombocytopenia in pregnancy
2. decreasing platelet count or newly discovered thrombocytopenia in pregnancy, which may or may not be related to pregnancy
3. acute onset of thrombocytopenia in the setting of severe pregnancy complications

#### Table 1. Causes and Relative Incidence of Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Category</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-existing thrombocytopenia</td>
<td>1-4%</td>
</tr>
<tr>
<td>Eclampsia</td>
<td>15-20%</td>
</tr>
<tr>
<td>Preeclampsia</td>
<td>2-15%</td>
</tr>
<tr>
<td>HELLP syndrome</td>
<td>0.6%</td>
</tr>
<tr>
<td>Drug-induced thrombocytopenia</td>
<td>1-4%</td>
</tr>
<tr>
<td>Secondary (ITP)</td>
<td>10-15%</td>
</tr>
</tbody>
</table>

#### Table 2. Basic Laboratory Evaluation of Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Test</th>
<th>Recommended/Not Recommended</th>
</tr>
</thead>
<tbody>
<tr>
<td>FDP</td>
<td>Recommended</td>
</tr>
<tr>
<td>Coagulation induced platelet aggregation test</td>
<td>Recommended</td>
</tr>
<tr>
<td>Bone marrow biopsy</td>
<td>Recommended</td>
</tr>
<tr>
<td>Direct antiglobulin (Coombs) test</td>
<td>Not recommended</td>
</tr>
<tr>
<td>VWD type IIB testing</td>
<td>Recommended</td>
</tr>
<tr>
<td>DIC testing</td>
<td>Recommended</td>
</tr>
<tr>
<td>Thyroid function tests</td>
<td>Recommended</td>
</tr>
<tr>
<td>Antiphospholipid antibodies</td>
<td>Recommended</td>
</tr>
<tr>
<td>Plasma kallikrein test</td>
<td>Recommended</td>
</tr>
</tbody>
</table>

#### Table 3. Therapeutic Options for Thrombocytopenia in Pregnancy

<table>
<thead>
<tr>
<th>Condition</th>
<th>First line therapy</th>
<th>Second line therapy</th>
<th>Third line therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary ITP (1-4%)</td>
<td>Danazol [X]</td>
<td>IVIg</td>
<td>Splenectomy (second trimester)</td>
</tr>
<tr>
<td>Secondary (ITP) (10-15%)</td>
<td>Cyclophosphamide [D]</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>HELLP syndrome</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Sepsis with systemic disorders</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Antiphospholipid antibodies</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Nitritoidal deficiency</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Bacterial sepsis</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>End-stage liver disease</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
<tr>
<td>Acute renal failure</td>
<td>TTP/HUS</td>
<td>Oral corticosteroids—initial response 2-14 days, peak response 2-7 days [C]</td>
<td></td>
</tr>
</tbody>
</table>


**A. Severe Preeclampsia**

1. Preeclampsia, which affects 5-8% of pregnant women, is diagnosed when:
   - a systolic BP ≥ 140 mm Hg or diastolic BP ≥ 90 mm Hg is present accompanied by proteinuria defined as urinary excretion of ≥ 0.3 g protein/24 hour or:
   - in a woman with previously normal blood pressure ≥ 150/100 mm Hg at term, while 125-120 mm Hg systolic and ≥ 80 mm Hg diastolic is the threshold. **

**B. HELLP Syndrome**

HELLP syndrome, which affects 0.6% of pregnant women, is a variant of preeclampsia and is defined as:
1. 15-20% of all women develop a platelet count < 100 x 10^9/L at term, while 125-120 mm Hg systolic and ≥ 80 mm Hg diastolic is the threshold. **
2. Eclampsia is new-onset Grand Mal seizures in a woman with preeclampsia.
3. Superimposed pre-eclampsia may develop in a woman with a history of chronic hypertension and is manifested by:
   - development of, or a sudden increase in hypertension after 20 weeks of gestation
   - a sudden increase in hypertension after 20 weeks gestation, or:
   - the development of the HELLP syndrome


**C. New-Onset of Thrombocytopenia in the Setting of Severe Preeclampsia**

In preeclampsia, thrombocytopenia is a common and potentially serious complication. The minimum platelet count for the placement of regional anesthesia is ≥ 50 x 10^9/L, while in preeclampsia, the minimum platelet count is ≥ 80 x 10^9/L. While platelet transfusion is not recommended in the management of primary immune thrombocytopenia, Blood 2010;115(2):168-186.

**V. Management of ITP at the Time of Delivery**

- Current recommendations aim for a platelet count of ≥ 50 x 10^9/L prior to labor and delivery as the risk of cesarean delivery is present with every labor.
- The minimum platelet count for the placement of regional anesthesia is ≥ 50 x 10^9/L, while in preeclampsia, the minimum platelet count is ≥ 80 x 10^9/L. While platelet transfusion is not recommended in the management of primary immune thrombocytopenia, Blood 2010;115(2):168-186.
- For a woman whose platelet count is ≥ 80 x 10^9/L but has not required therapy during pregnancy, oral prednisone (or prednisolone) can be started 10 days prior to anticipated delivery at a dose of 10-20 mg daily and titrated as necessary.

- Given the difficulty predicting severe thrombocytopenia in monkeys and the very low risk of intracranial hemorrhage (<1% or 0.5%) or mortality (<1%), the platelet count should be determined by objective indication.

- Perinatal umbilical blood sampling (PBS) or fetal scalp blood sampling is not helpful in predicting neonatal thrombocytopenia, or the very low risk of intracranial hemorrhage (<1% or 0.5%).

- The infant nadir platelet count occurs 2-5 days after delivery and a spontaneous rise occurs by day 7.

- Women with ITP are at an increased risk of venous thrombembolism, and as no form of prophylaxis for postpartum thrombocytopenia, Blood 2011;121(1):38-47.


**VI. Acute Onset of Thrombocytopenia in the Setting of Preeclampsia**

"The American Society of Hematology 2011 Evidence-Based Practice Guidelines for Immune Thrombocytopenia" recommends a starting dose of prednisone of 1mg/kg daily, there is no evidence that a higher starting dose is better than a lower dose. Other experts recommend a dosage of starting dose of 0.25 to 0.5 mg/kg daily.

- Medications are adjusted to maintain a safe platelet count (below).

Antihypertensives are used to control blood pressure.

If the patient is <34 weeks gestation and maternal and fetal status are not reassuring, the patient should be delivered as soon as she is able. Specific features predictive of poor postpartum outcome are when plasma exchange is otherwise contraindicated.

VII. Thrombotic Thrombocytopenia Purpura (TTP)/Atypical Hemolytic Uremic Syndrome (aHUS)

Thrombotic thrombocytopenia purpura (TTP) and atypical hemolytic uremic syndrome (aHUS) are rare but serious conditions of the third trimester (1 in 20,000 pregnancies). They differ from the HELLP syndrome by various features yet share characteristics with it.

While the use of eculizumab has been described in paroxysmal nocturnal hemoglobinuria during pregnancy, as yet no reports exist on eculizumab in aHUS during pregnancy.

The therapeutic plasma exchange is generally used to treat thrombocytopenia and hemolysis when other therapies have failed to improve thrombocytopathy.

Therapeutic plasma exchange— if thrombocytopenia, hemolysis or renal failure continues to worsen after 48–72 hours postpartum, therapeutic plasma exchange should be repeated as appropriate. Special considerations for TTP/aHUS precipitated by pregnancy may be difficult (see Table 6).

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Antihypertensives are used to control blood pressure.

Magnesium sulfate is used to prevent seizures.

1. Obstetric management

- Cồn-gradually—may be the platelet count and other laboratory parameters more quickly, but have not been shown to improve long-term maternal or fetal outcomes.
- Supportive care with blood products—there is no contraindication to platelet transfusion.
- Therapeutic plasma exchange—therapy of choice for HELLP syndrome, hemolysis or renal failure continues to worsen 48-72 hours postpartum

Elevated uric acid

Coagulopathy

Abdominal pain

Ascites or bright liver on ultrasound scan

Vomiting

Leukocytosis

Thrombocytopenia

Hemolysis

Abnormal peripheral smear

Elevated liver enzymes

LFTs, CR, and metalloproteinase with thrombospondin type 1 motif, member 13

AFLP has overlapping features with HELLP, but there is no well-established definition of the condition that clearly differentiates it from HELLP.

C. Acute Fatty Liver of Pregnancy (AFLP)

- AFLP is a rare but serious condition of the third trimester (1 in 20,000 pregnancies).

D. Management of Severe Preeclampsia, the HELLP Syndrome, or AFLP with a Prospective National Study of Acute Fatty Liver of Pregnancy in the UK. Gut. Reference: Knight M, Nelson-Piercy C, Kurinczuk JJ, Spark P, Brocklehurst P.

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Magnesium sulfate is used to prevent seizures. If the patient is <34 weeks gestation and maternal and fetal status are A prospective national study of acute fatty liver of pregnancy in the UK. Gut.

A prospective study from Southwest Wales (the "Swansea Criteria") and have established definition of the condition that clearly differentiates it from subsequent been used in other studies. Six or more of these criteria in the absence of another explanation were required for a diagnosis of AFLP.

Table 5. The Swansea Criteria for the Diagnosis of AFLP—6 Necessary

<table>
<thead>
<tr>
<th>Disease</th>
<th>Criteria</th>
<th>Percentage</th>
<th>Incidence</th>
<th>Prevalence</th>
<th>Screening</th>
<th>Diagnosis</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>HELLP syndrome</td>
<td>&lt;10% of women</td>
<td>70% of cases</td>
<td>&lt;5% of cases</td>
<td>&lt;5% of cases</td>
<td>&lt;15%</td>
<td>Elevated LFTs</td>
<td>Therapeutic plasma exchange</td>
</tr>
<tr>
<td>AFLP</td>
<td>&gt;30%</td>
<td>&gt;1%</td>
<td>&lt;1%</td>
<td>&gt;1%</td>
<td>&lt;15%</td>
<td>Elevated LFTs</td>
<td>Therapeutic plasma exchange</td>
</tr>
</tbody>
</table>


VI. Thrombotic Thrombocytopenia Purpura (TTP)/Atypical Hemolytic Uremic Syndrome (AHUS)

Differential diagnosis between severe preeclampsia, HELLP, AFLP, or evolving TTP/AHUS if platted by pregnancy may be difficult (see Table 5). While the use of ecclsumol is has been described in paroxysmal nocturnal hemoglobinuria during pregnancy, as yet no reports exist on ecclsumol in AHUS during pregnancy.

Since therapeutic plasma exchange has been shown to improve the outcome of all of these conditions, when plasma exchange is otherwise indicated, diagnostic certainty is not required.

Table 5. Selected Causes of Thrombocytopenia During Pregnancy

<table>
<thead>
<tr>
<th>Disease</th>
<th>Incidence</th>
<th>Diagnosis</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gestational thrombocytopenia</td>
<td>&lt;5%</td>
<td>Elevated platelet count</td>
<td>Therapeutic plasma exchange</td>
</tr>
<tr>
<td>TTP</td>
<td>&lt;1%</td>
<td>Elevated platelet count</td>
<td>Therapeutic plasma exchange</td>
</tr>
</tbody>
</table>


This guide is intended to provide the practitioner with clear principles and strategies for quality patient care and does not establish a fixed set of rules that practitioners may follow. For further information, contact the ASH Department of Government Relations, Practice, and Scientific Affairs at 202-776-0444. ASH website: www.hematology.org/practiceguidelines © 2013 by the American Society of Hematology. All rights reserved.