Severe Idiopathic Thrombocytopenia Purpura

Definition: Idiopathic Thrombocytopenia Purpura (ITP) is an autoimmune blood disorder characterised by reduction of platelets levels in blood. Normal platelet level is 150-450 x 10^9/L. In severe ITP the platelet count is below 20 x 10^9/L and is life threatening.

Etiology: Unknown, hence the name ‘idiopathic’. However, viral infections, such as chicken pox, rubella, mumps and live vaccinations may trigger ITP in children. The highest risk group are children between the ages of 2-5 years.

Pathophysiology
Normal bone marrow contains two types of cells, megakaryocytes and red bone marrow cells. The megakaryocytes are the precursors of the platelets responsible for clotting. The normal life span of platelets is between 3-10 days. The spleen is responsible for destruction of old platelets as well as affected platelets.

For unknown reasons, antibodies are triggered to attack either of the megakaryocytes in the bone marrow or the already produced platelets. The megakaryocytes become immature and less productive. When the platelets are attacked by the antibodies, the spleen recognises them as foreign bodies and destroys them. The destruction of platelets occurs very rapidly, leading to reduced platelet levels.

Signs and Symptoms
- Petechiae or purpura
- Ecchymosis (Bruises)
- Persistent bleeding from cuts
- Mucosal/Gingival bleeding
- Frequent or heavy nose bleeding
- Haematuria
- Splenomegaly

Complications
- Intracranial/ cerebral haemorrhage
- Bleeding into diaphragm can lead to pulmonary complications.
- Gastrointestinal bleeding
- Anaemia
- Hematomas on nerve or brain tissue can lead to paralysis.
- Purpural lesions can occur in vital organs such as brain, kidneys and intestinal tract, compromising their functions.
- Hypothyroidism or hyperthyroidism, as antibodies attack the thyroid gland.

Nursing Care
Mother-child interaction:
- Explain the condition and treatment to the caregiver to reduce anxiety. She can then reassure the child.
- Allow the caregiver to be with the child to promote interaction.
- Pain/comfort management:
  - Assess for pain and manage accordingly.
  - Promote sleep. Nurse child in a comfortable position.
Nutrition:
- Encourage the child to eat dark green leafy vegetables. They promote clotting.

Hydration:
- Observe and report the amount and location of bleeding episodes
- Secure IV line for fluids, medication and possibly blood transfusions.
- Exert pressure on any puncture sites for 5 to 10 minutes
- Monitor fluid balance by recording input and output daily.
- Describe and monitor urine and vomiting for blood
- Transfuse blood if there are signs of cardiac failure such as tachypnoea

Prevent infection:
- Apply infection prevention measures such as hand washing, linen changes and sterility during procedures.
- Encourage short stay to prevent nosocomial infections
- Maintain skin and mucosal integrity:
  - Provide primary care such as bathing, frequent nappy changes, mouth care and skin care to promote skin and mucosal integrity
  - Use a soft toothbrush to prevent gum mucosal breakdown
  - Protect area of petechiae from further injuries
- Promote exercises as tolerated

Support regulatory system:
- Monitor and record vital signs such as temperature, heart rate, respiratory rate and blood pressure hourly and report any deterioration
- Observe for signs of respiratory distress and cardiac failure, such as chest recessions, cyanosis and nasal flaring
- Maintain patient on strict bed rest during active bleeding episodes
- Obtain blood samples for haemoglobin levels and platelet count daily
- Avoid taking drugs like aspirin, because it impairs coagulation
- Monitor glucose levels daily.

Treatment
- Might not be required, depending on severity
- IV immune globulin (IVIG) or Rh immune globulin to prevent further destruction of platelets
- Medications (e.g. Etilrombopag/ Romiplostim) to increase platelet count
- Surgery to remove spleen, if site of destruction of platelets.

References
4. Joseph, T. Personal communication [2013, April, 21]