Polycythemia/Hyperviscosity

INTRODUCTION: Polycythemia, defined as a **venous hematocrit (Hct) >65%,** occurs in about 4% of term infants, and is more common with SGA (IUGR), LGA, infants of diabetic mothers, trisomy 21, twin-twin transfusion, Beckwith-Wiedemann syndrome and otherwise normal infants who receive an excessive placental transfusion.

PATHOPHYSIOLOGY:

1. Hct is the main determinant of blood viscosity, which is also affected by plasma proteins, especially fibrinogen. As Hct increases, viscosity increases, elevating vascular resistance and decreasing blood flow at any given perfusion pressure. As Hct rises above 60%, viscosity tends to increase exponentially. The effects of increased Hct on viscosity are greater in the pulmonary circulation; in experimental animals, pulmonary vascular resistance exceeds systemic vascular resistance when Hct >70%.

2. Clinically useful methods of measuring viscosity are currently not available. Therefore, **Hct is used as a proxy for viscosity**. Because viscosity is affected by other factors (*e.g.*, flow rate in small vessels, plasma protein concentrations, acidosis, red blood cell deformability), viscosity does not correlate directly with Hct.

3. Increased viscosity results in slowing of blood flow and sludging of red blood cells. As blood flow is further reduced, occlusion of small vessels may result in ischemia and consumption of platelets. Because glucose is transported mainly in plasma and relative plasma volume is decreased, the decreased blood flow can result in hypoglycemia.

CLINICAL FINDINGS depend on organ system affected, as indicated below:

Organ system	Signs & Symptoms
Skin	Plethora, prolonged capillary filling time
CNS - Mild	Lethargy, irritability, tremors, abnormal cry
CNS – Severe	Apnea, seizures, hypotonia
Pulmonary	Respiratory distress, cyanosis, persistent pulmonary hypertension
Renal	Hematuria, oliguria/ anuria, renal vein thrombosis
Hematologic	Thrombocytopenia, or other signs consistent with DIC
Gastro-intestinal	Abdominal distension, blood in stools, necrotizing enterocolitis
Metabolic	Persistent hypoglycemia

HEMATOCRIT SCREENING:
   •Asymptomatic infants with no risk factors: Hct measurement is not necessary.
   •Measure Hct in:
       -Any infant at risk for polycythemia (see Introduction above)
       -Any infant with symptoms that may be due to polycythemia

HEMATOCRIT SOURCE: Hct varies with source of blood. Capillary Hct (heel stick) is 5-15% >venous. Arterial Hct averages 6% <venous. Decisions regarding treatment of polycythemia should be made on the basis of **venous Hct**.

TREATMENT OF POLYCYTHEMIA is to reduce Hct by **partial exchange transfusion (Partial ExTx) with a plasma substitute**. **Do not perform phlebotomy,** which decreases blood volume, reduces blood pressure, and actually increases viscosity as blood flow rate decreases. Perform Partial ExTx for the following situations:
- Hct >65% in a symptomatic infant
- Hct >70% in an asymptomatic infant
- Consider partial exchange transfusion in symptomatic infants with Hct 60-65%

PARTIAL EXCHANGE TRANSFUSION (Partial ExTx):

1. Volume to be exchanged is calculated by the following equation:

$$\text{Volume (mL)} = \frac{\text{Initial Hct} - \text{Desired Hct}}{\text{Initial Hct}} \times \text{Weight (kg)} \times 90 \text{ mL/kg}$$

-Desired Hct should be 50 to 55%.
-Hypervolemia is common in polycythemia; use 90 mL/kg as estimated blood volume.
-Volume to be exchanged in a term infant is almost always in the range of 40-60 mL/kg. If calculated volume is outside this range, re-check the calculations.

2. Use 0.9% NaCl (isotonic saline) for Partial ExTx. It effectively maintains lowered Hct. Fresh Frozen Plasma and 5% albumin contain proteins that may add to viscosity.

3. As soon as the decision has been made to lower the Hct, obtain **informed consent** from parents. Risks of polycythemia/hyperviscosity include cerebrovascular accidents, renal vein thrombosis, hypoglycemia, necrotizing enterocolitis and jaundice. Those outweigh the potential risks of Partial ExTx (thrombosis, infection, vascular perforation, limb ischemia, hemorrhage), which are rare with Partial ExTx.

3. Technique of Partial ExTx: Perform Partial ExTx as soon as decision has been made to lower Hct. Use **aliquots of 5 mL/kg**; withdraw blood first, then infuse an equal amount of saline. Routes of Partial ExTx are, in decreasing order of preference:

-**Umbilical arterial catheter (UAC):** Insert UAC in proper position (tip in descending aorta, below 3rd lumbar vertebra). Obtain radiograph (chest and abdomen) to ensure proper position. If patient has cardio-respiratory distress, consider using UAC for further monitoring; otherwise, remove UAC after completion of Partial ExTx.

-**Umbilical venous catheter (UVC):** This can be done in one of two ways:
   (a) Insert UVC only as far as needed to withdraw blood. Then, while simultaneously infusing an equal volume of saline through a peripheral vein, withdraw calculated volume of blood from UVC. UVC may need frequent flushing with 1-2 mL of heparinized 0.9% NaCl.
   (b) Insert UVC so tip is in right atrium (See section on Intravascular Catheters, P. 25). Use push-pull technique. Remove UVC at end of procedure.

-**Peripheral arterial cannula:** Use this for blood withdrawal and peripheral IV for simultaneous infusion of saline. This method is theoretically the safest. However, due to technical difficulties, it often prolongs the procedure unnecessarily.

4. For all umbilical catheters (arterial and venous), measure blood pressure, pH and blood gas tensions. At completion of procedure (any route), measure Hct and platelet count.

5. Monitor vital signs throughout procedure and observe for catheter related problems. If these occur, discontinue procedure and remove catheter.

6. After Partial ExTx, **keep infant NPO for at least 4h**. Give IV glucose water to prevent hypoglycemia. **If thrombocytopenia is present, keep infant NPO until platelet count is in normal range.** Repeat Hct measurement 4h after procedure.