

POLYCYTHAEMIA • 1/2

RECOGNITION AND ASSESSMENT

Definition

- Peripheral **venous** haematocrit (Hct) >65%
- Symptoms rarely occur with peripheral Hct of <70%
- Hct peaks at 2 hr after birth and then decreases with significant changes occurring by 6 hr
- **Common in the newborn and affects approximately between 1 to 5% of babies**

Clinical consequences

- Hyperviscosity
- Decreased blood flow and impaired tissue perfusion
- Thrombus formation

Complications

- Cerebral micro-infarction and adverse neurodevelopmental outcome
- Renal vein thrombosis
- Necrotising enterocolitis (NEC)
- **Platelet consumption**

Causes

Intra-uterine increased erythropoiesis	Erythrocyte transfusion
<ul style="list-style-type: none">• Placental insufficiency (SGA)• Postmaturity• Maternal diabetes• Maternal smoking• Chromosomal abnormalities: trisomy 21, 18, 13• Beckwith–Wiedemann syndrome• Congenital adrenal hyperplasia• Neonatal thyrotoxicosis• Congenital hypothyroidism	<ul style="list-style-type: none">• Maternal-fetal• Twin-to-twin transfusion• Delayed cord clamping• Unattended delivery

Symptoms and signs

Commonly plethoric but asymptomatic. The symptoms evolve over the first 24 hours as the haematocrit rises with the physiological decrease in plasma volume. The infants appear plethoric and can become cyanosed, particularly when active.

Cardiorespiratory	<ul style="list-style-type: none">• Respiratory distress• Tachycardia• Persistent pulmonary hypertension of the newborn• Congestive cardiac failure
CNS	<ul style="list-style-type: none">• Lethargy, hypotonia within 6 hr• Difficult arousal, irritability• Jittery• Easily startled• Seizures
GIT	<ul style="list-style-type: none">• Poor feeding• Vomiting• NEC
Metabolic	<ul style="list-style-type: none">• Hypoglycaemia• Hypocalcaemia• Jaundice
Haematological	<ul style="list-style-type: none">• Thrombocytopenia
Renal	<ul style="list-style-type: none">• Renal vein thrombosis• Renal failure

Before making a diagnosis of polycythaemia, it is important to exclude dehydration. Re-weigh the baby to look for excessive weight loss. Examine for signs of dehydration. Check urea and electrolytes. If present, correct by increasing fluid intake. Measure the haematocrit again after correction of dehydration.

INVESTIGATIONS

In all unwell babies and at-risk babies who look plethoric (as mentioned above)

- FBC/Hct
- If Hct >65%, repeat a free-flowing venous sample or obtain arterial Hct (capillary Hct sample unreliable)
- If polycythaemic, check blood glucose and serum calcium

IMMEDIATE TREATMENT

Asymptomatic babies with Hct 65-70%

- Observe closely and monitor for common complications such as hypoglycaemia and hyperbilirubinaemia.
- Monitor oral intake, body weight and urine output
- Repeat venous haematocrit in 12 hours. If it remains less than 70% and the infant remains asymptomatic, continue this approach for 24 hours and recheck haematocrit.

Asymptomatic babies with Hct >70%

- Ensure liberal intravenous fluid intake 1 day ahead (see **Intravenous fluid therapy** guideline)
- Check urea and electrolytes and bilirubin regularly and start maintenance sodium chloride (and potassium chloride) additives at 24-48 hours of life to avoid iatrogenic hyponatraemia/ hypokalaemia.
- Repeat venous Hct after 6 hr
- if still high, discuss with consultant to consider partial exchange transfusion

Symptomatic babies with Hct >65%

- Possible symptoms: fits and excessive jitteriness, with neurological signs and refractory hypoglycaemia

Treatment

- Dilutional exchange transfusion. Discuss with consultant due to increased risk of NEC
- use of haemodilution (partial exchange transfusion) for treatment of polycythaemia is not supported by evidence and treatment of asymptomatic babies is not recommended
- explain to parents need for exchange and possible risks before performing dilutional exchange transfusion. Partial exchange transfusion increases risk of NEC
- **Isovolumetric PET reduces the haematocrit without causing hypovolaemia. The procedure reverses the reduction in cerebral blood flow, cardiac index and oxygen transport attributed to hyperviscosity. However, PET does not appear to affect long term outcome. The long term outcome is more likely to be related to the underlying cause of polycythaemia**
- use sodium chloride 0.9% (see **Exchange transfusion** guideline)
 - Volume to be exchanged generally = **20 mL/kg over 30mins** (this usually reduces the haematocrit to below 60%).
 - **or use following formula:**

$$\frac{\text{blood volume} \times (\text{observed Hct} - \text{desired Hct})}{\text{observed Hct}}$$

* blood volume estimated 80–90 mL/kg in term babies, 90–100 mL/kg in preterm Babies

Procedure

1. Insert peripheral venous catheter and peripheral arterial catheter or insert umbilical venous catheter.
2. Start infusion of 20mls/kg 0.9% saline over 30 minutes via peripheral venous catheter.
3. Remove 5-10 mls aliquots of blood via peripheral arterial catheter (or UVC if using).
4. Continue removing 5-10 ml aliquots until a total of 20 ml/kg has been exchanged over 30 minutes.
5. Recheck FBC and venous haematocrit at end of procedure.
6. Remove lines if no longer required.

Continuous nursing observation will be required during the exchange transfusion

SUBSEQUENT MANAGEMENT

- Babies who required dilutional exchange transfusion require long-term neurodevelopmental follow-up
- Otherwise, follow-up will be dependent on background problem