POLYCYthaemia 2011-13

POLYCYthaEMIA

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

Clinical consequences
- Hyperviscosity
- Decreased blood flow and impaired tissue perfusion
- Microthrombus formation

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

Causes

<table>
<thead>
<tr>
<th>Intra-uterine increased erythropoiesis</th>
<th>Erythrocyte transfusion</th>
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<tbody>
<tr>
<td>Placental insufficiency (SGA)</td>
<td>Maternal-fetal</td>
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<tr>
<td>Postmaturity</td>
<td>Twin-to-twin transfusion</td>
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<td>Maternal diabetes</td>
<td>Delayed cord clamping</td>
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<td>Maternal smoking</td>
<td>Unattended delivery</td>
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<tr>
<td>Chromosomal abnormalities: trisomy 21, 18, 13</td>
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<tr>
<td>Beckwith–Wiedemann syndrome</td>
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<td>Congenital adrenal hyperplasia</td>
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<tr>
<td>Neonatal thyrotoxicosis</td>
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<td>Congenital hypothyroidism</td>
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Symptoms and signs
- Commonly plethoric but asymptomatic

Cardiorespiratory
- Respiratory distress
- Persistent pulmonary hypertension of the newborn (PPHN)
- Congestive cardiac failure

CNS
- Lethargy, hypotonia within 6 hr
- Difficult arousal, irritability
- Jittery
- Easily startled
- Seizures

GIT
- Poor feeding
- Vomiting
- NEC

Metabolic
- Hypoglycaemia
- Hypocalcaemia
- Jaundice

Haematological
- Thrombocytopenia

Renal
- Renal vein thrombosis
- Renal failure

INVESTIGATIONS
In all unwell infants and at-risk infants who look plethoric (as mentioned above)
- FBC/Hct
- If Hct >65%, repeat using a 21 gauge needle to check that venous blood flows freely, or obtain arterial Hct
- If polycythaemic, check blood glucose and serum calcium
IMMEDIATE TREATMENT

- Ensure infants at risk have liberal fluid intake one day ahead (e.g. 90 mL/kg/day on day 1)

Asymptomatic infants with Hct >70%

- Repeat venous Hct after 6 hr
- if still high, discuss with consultant (current evidence does not show any benefit in treating asymptomatic babies)

Symptomatic babies with Hct >65%

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

Treatment

- Dilutional exchange transfusion. Discuss with consultant
- explain need for exchange and possible risks to parents before performing dilutional exchange transfusion. Partial exchange transfusion increases risk of NEC
- use sodium chloride 0.9% – see Exchange transfusion guideline
- Volume to be exchanged = 20 mL/kg
- Perform exchange via umbilical venous catheter (UVC) or via peripheral arterial and IV lines
- Take 5–10 mL aliquots and complete procedure over 15–20 min

SUBSEQUENT MANAGEMENT

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