CLINICAL PRACTICE GUIDELINE

Neonatal screening: haemoglobin disorders

This document should be read in conjunction with the [Disclaimer](#)

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Testing for neonatal significant haemoglobin disorders

Aim
Appropriate collection of cord blood samples for neonates identified as at risk of a significant haemoglobin disorder (other than sickle cell disease which is covered later in this guidance).

Background Information
KEMH has an established process of screening women who may be deemed at risk of carrying an inherited haemoglobin disorder in the antenatal period:

- To assess the risk of a couple having a severely affected child, and
- To enable informed choice surrounding decision making.

For further information refer to Clinical Practice Guideline: Antenatal Care: Haemoglobinopathy Screening in Pregnancy

If the fetus is determined to be at risk of the following significant haemoglobin disorders:

- Beta thalassaemia major
- Haemoglobin H disease
- Haemoglobin E/Beta thalassaemia

A completed ‘Haemoglobinopathy Screen’ sticker on the Obstetric Special Instruction sheet (MR 004) will direct neonatal testing requirements (if required). In addition the maternal medical records may include a Neonatal Management Plan/Folder which will identify additional and specific neonatal management plans for the newly delivered infant.
Procedure

1. If a sample is required for haemoglobinopathy testing, collect an extra cord EDTA sample (minimum sample required is 0.5mL). NB: the cord EDTA sample sent to Blood Bank cannot be used for haemoglobinopathy testing.

2. If no cord sample is available collect a peripheral blood sample (capillary blood collection – heel stab). See Clinical Guideline: O&M, Neonatal Care: Capillary Blood Collection

3. Label the sample with the neonate’s details.

4. Document in the maternal notes and neonatal history chart (MR 410) that a sample for haemoglobinopathy testing has been collected.

5. Use the CPOE ‘down-time’ Pathology request form (yellow) with the “Neonatal Haemoglobinopathy Screening” sticker and tick the “Exclude other significant Haemoglobinopathy” box.

6. The form must be completed by a member of the Paediatric, Haematology or Genetic Department. Specify the maternal and paternal globin gene abnormalities (genotype) if this information is available. This is to assist in directing appropriate laboratory testing.

7. Send the specimen to specimen collection in pathology.

8. If the haemoglobin studies results indicate a significant haemoglobinopathy, Pathwest Laboratory Medicine will notify the Princess Margaret/Perth Children's Hospital Haematology Department and they will make the necessary arrangements to review the neonate. This notification will be indicated on the haemoglobin studies report.
Cord Blood Screening Program: sickle cell disease

Aim
To identify neonates born at KEMH with Sickle Cell Disease so that treatment can be commenced as early in life, as possible, in order to optimise outcomes.

Key points
Cord blood for Sickle Cell Disease screening is collected from neonates born at KEMH to women of indigenous African origin with:
- Unknown sickle cell status or
- Sickle cell trait / sickle cell disease.

Background
- Sickle Cell Disease is an autosomal recessive disorder which implies that it must be inherited through both parents who may have the disorder themselves or be carriers.
- Sickle Cell Disease occurs when abnormal genes are inherited from both parents.
- Sickle Cell trait is when a person inherits only one sickle cell gene and does not have the disease.
- A patient information sheet on sickle cell anaemia is available at https://patient.info/health/sickle-cell-disease-sickle-cell-anaemia
- Indigenous Africans are the ethnic population with the highest prevalence of sickle cell disease and this is the reason why babies born to these women have been chosen as the target population for this screening program.
- Due to changes in migration patterns, sickle cell disease also occurs in women of Afro-American, Afro-Caribbean and Central/South American origin. See Clinical Guideline, O&M: Antenatal Care: Haemoglobinopathy Screening and Referral.

Procedure
For women of indigenous African origin
1. Check the woman’s medical record or isoft Clinical Manager (iCM) for a haemoglobin studies report regarding sickle cell status.
2. If there are no results available, or the results state haemoglobin S (sickle cell) trait, or Haemoglobin S (sickle) disease, testing is required to exclude neonatal sickle cell disease.
3. If testing is required, collect an extra cord blood EDTA sample (minimum sample required is 0.5mL). NB: the cord EDTA sample sent to the Blood Bank cannot be used for haemoglobinopathy testing.
4. If no cord sample is available collect a peripheral blood sample (heel stab) as per Clinical Guideline, O&M, Neonatal Care: Capillary Blood Collection.
5. Label the sample with the neonate’s details.
6. Use a CPOE ‘down-time’ Pathology Request form (yellow) with the ‘Neonatal Haemoglobinopathy Screening’ sticker and tick the ‘Exclude Sickle cell Disease’ box.

![Copy of Results: KEMH MFM Service](image)

**NEONATAL HAEMOGLOBINOPATHY SCREENING**

**TICK INDICATION FOR TESTING**

- [ ] Exclude Sickle Cell Disease
- [ ] Exclude other significant Haemoglobinopathy
  
  (Parental genotype to be included to direct testing)

**Maternal genotype:**

**Paternal Details:**
- Name:
- DOB:
- Genotype:

7. A midwife may sign the request form.
8. The midwife shall document in the maternal notes and in the neonatal history chart MR410 that the Sickle cell Disease Screen has been collected.
9. Send the specimen to specimen collection in pathology.
10. If the haemoglobin study results indicate Sickle cell Disease, Pathwest Laboratory Medicine will directly notify the Princess Margaret/Perth Children’s Hospital Haematology Department and they will make the necessary arrangement to review the neonate. This notification will be indicated on the haemoglobin studies report.
Cord Blood for Haemoglobinopathy Screening

Aim
To facilitate neonatal diagnosis of sickle cell disease by the collection of cord blood from neonates whose parents are both of indigenous African origin, or where one parent is known to have sickle cell trait and the other has not been screened.

Key points
- Cord blood for haemoglobinopathy screening is only collected from babies where both parents are of indigenous African origin or where one parent is known to have sickle cell trait and the other has not been screened (regardless of ethnicity).
- NB: Routine screening is directed ONLY at sickle cell disease.
- Cord blood for haemoglobinopathy screening is only collected from babies born at KEMH.

Procedure
1. Following the birth of the baby place 0.5-1.0mL of cord blood in an EDTA tube.
2. Label appropriately with the neonates details.
3. Complete a pathology request form – state “Haemoglobin studies – exclude sickle cell disease’ on the form. Midwives may sign the request form for sickle cell only.
4. Place the specimen and completed request form in the utility room for collection.

References and resources

Related policies
Nil

Related WNHS policies, procedures and guidelines
KEMH Clinical Guidelines, Obstetrics & Midwifery::
- Antenatal Care: Haemoglobinopathy Screening and Referral
- Neonatal Care: Capillary Blood Collection
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