Aneurin Bevan University Health Board

Sickle Cell Anaemia and Haemoglobinopathy Screening and Management in Pregnancy Guidelines

N.B. Staff should be discouraged from printing this document. This is to avoid the risk of out of date printed versions of the document. The Intranet should be referred to for the current version of the document.

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Approved by: Maternity Clinical Effectiveness Forum | Review by: 14 September 2020
Owner: Maternity Services | Policy No: ABUHB/F&T/0713

Contents:

1. Executive Summary:

This document should act as guidelines for the screening for sickle cell anaemia and haemoglobinopathies and the management women with sickle cell anaemia in pregnancy and postpartum period. The opinion expressed in the guideline are evidence and reflects professional opinion. It is designed to support safe and effective practice.

1.1 Scope of the guidelines:

- The guideline applies to all clinicians working within the maternity services.

1.2 Essential implementation criteria:

- Auditable standards are stated.

2. Aims

- To provide support to clinical decision making
- To provide support for evidence based management

3. Responsibilities

- The maternity management team

4. Training

- Staff are expected to access appropriate training where provided
- Training needs will be identified through appraisal and clinical supervision

5. Monitoring and Effectiveness:

- Local service improvement plan will guide monitoring and effectiveness. This policy has undergone an equality impact assessment screening process using toolkit designed by NHS centre Equality and Human rights.
- Details of the screening process for this policy are available from the policy owner.

6. Implementation

- The guidelines will be implemented for the screening of women for sickle cell anaemia and haemoglobinopathies and to guide the

management of women with sickle cell anaemia in pregnancy and postpartum period.

7. Standards for Health Services Wales

Has an equality impact assessment been carried out?

- YES

Has any adverse impact been identified?

- NO

8. Environmental Impact

- NO

9. Audit

- Audit tools have been incorporated in the protocol.

10. Review

- Protocol to be reviewed in 3 years.

11. Appendices

Appendix 1 Protocol

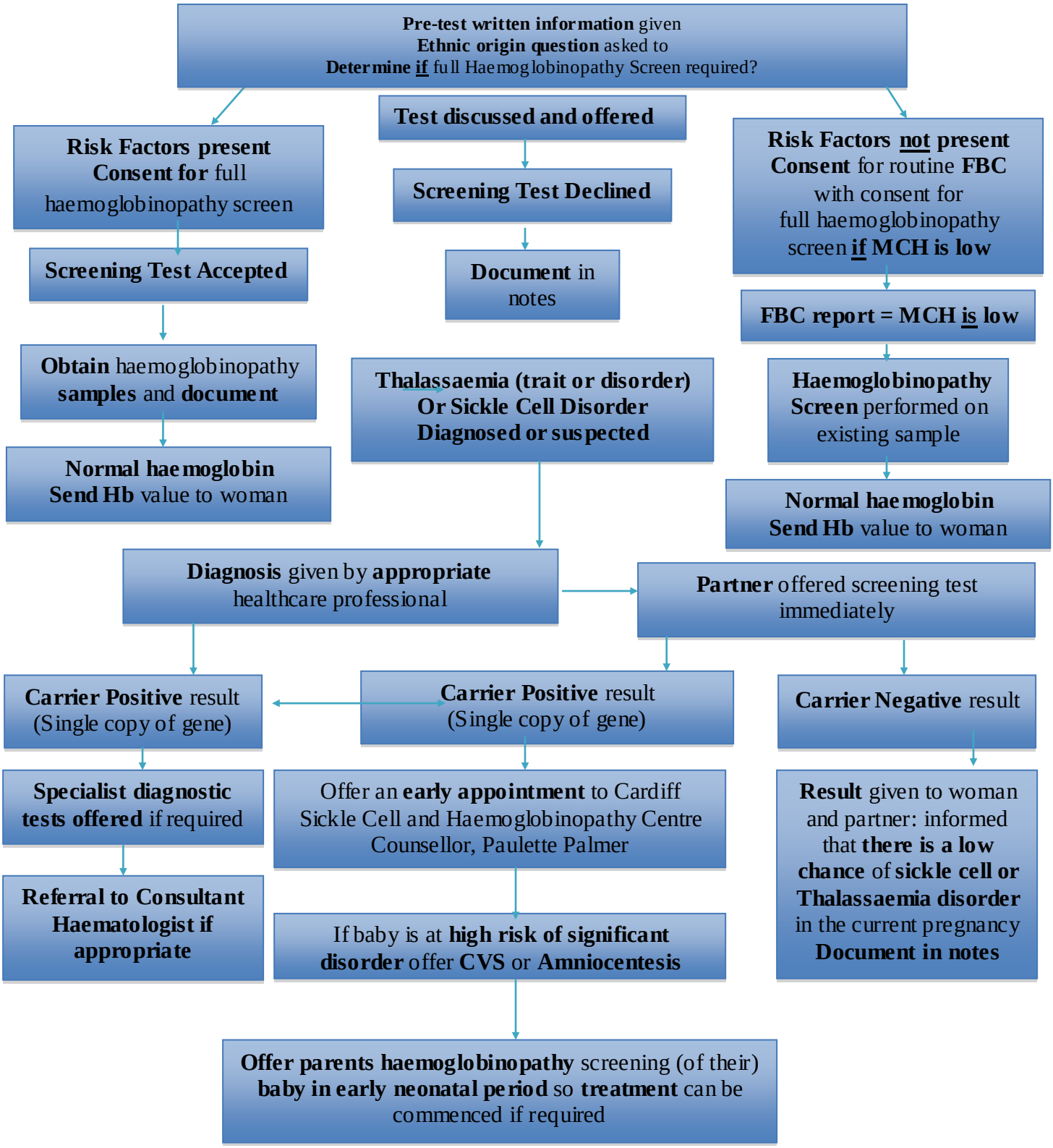
Sickle Cell Disease/Anaemia (SCD)

- Most common inherited condition worldwide
- In the UK there are between 12,000 and 15,000 affected individuals with SCD
- Each year, approximately 100–200 pregnancies in women with SCD.
- Most prevalent in individuals of African descent
- Autosomal recessive disorder

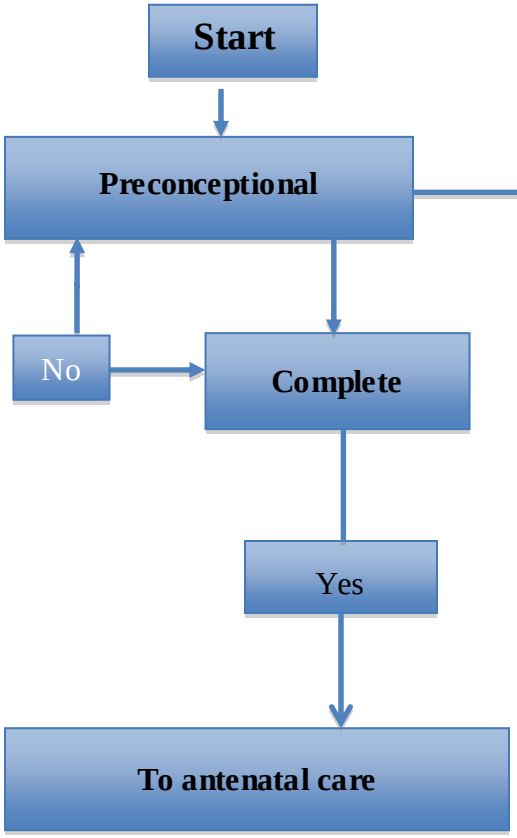
Clinical Features

- Chronic Anaemia
- Painful sickle crisis
- Acute chest syndrome
- Endocrine - delayed pubertal development, gonadal failure, diabetes and primary hypothyroidism
- Splenic infarction
- Chronic organ damage
- Neurological complications such as stroke or silent infarction

Sickle Cell and Thalassaemia Screening



Pre Conceptional Advice and Work up



Genetic screening

Haemoglobinopathy status of partner, If positive – counselling as per National screening Guidelines

Chronic disease surveillance - Annual screening

- ECG – screen for pulmonary HTN
- Retinal screening
- T2 Cardiac MRI for Iron overload
- U & E, LFT, Serum Ferritin level, Red cell antibodies

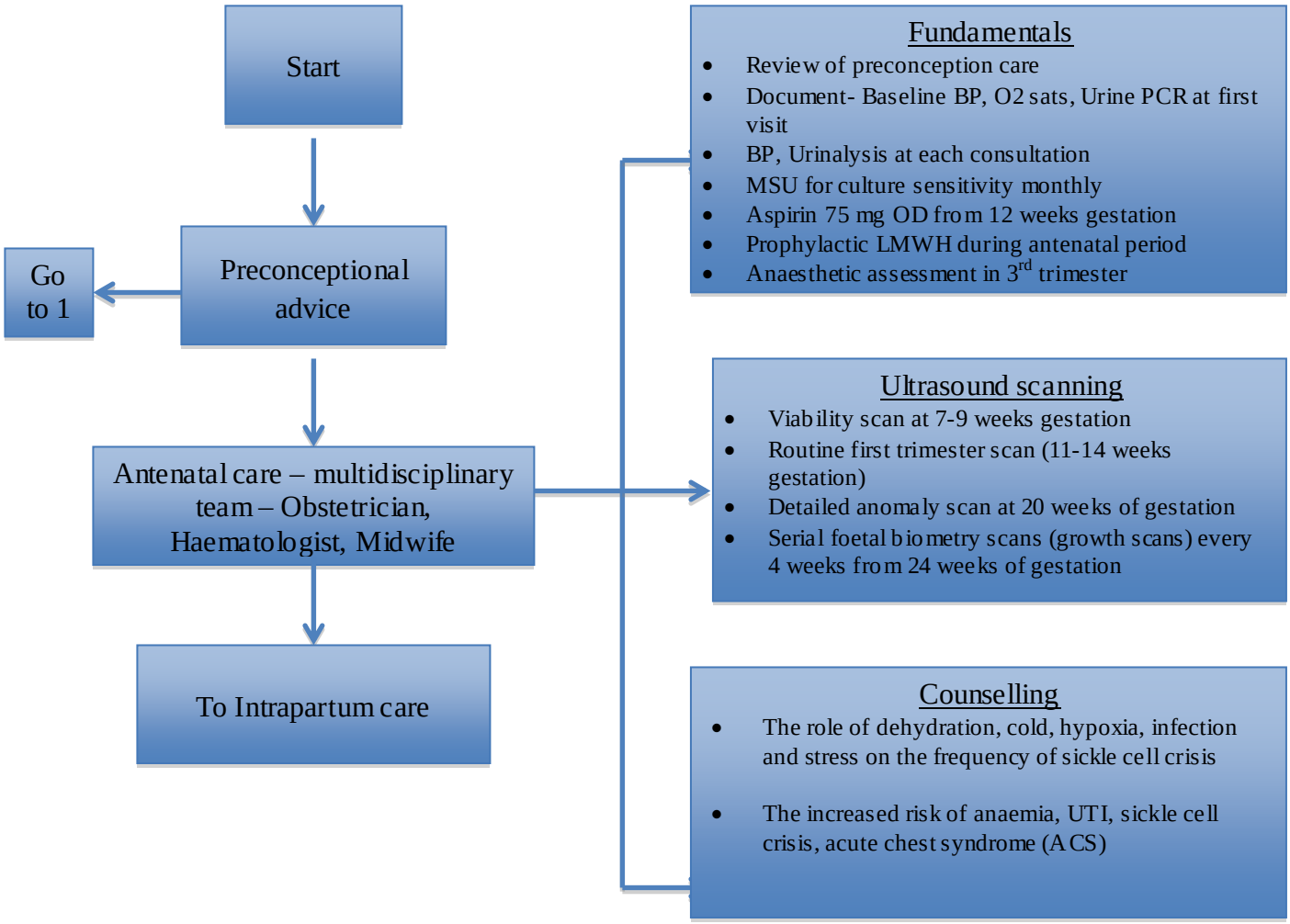
Medications

- Folic acid 5mg and Vit D 10 mcg once a day
- Phenoxy methylpenicillin 500mg bd prophylaxis (Erythromycin if allergic)
- Hydroxyurea – stop 3 months before conception
- ACE Inhibitors, Angiotensin receptor blockers – Stop before conception

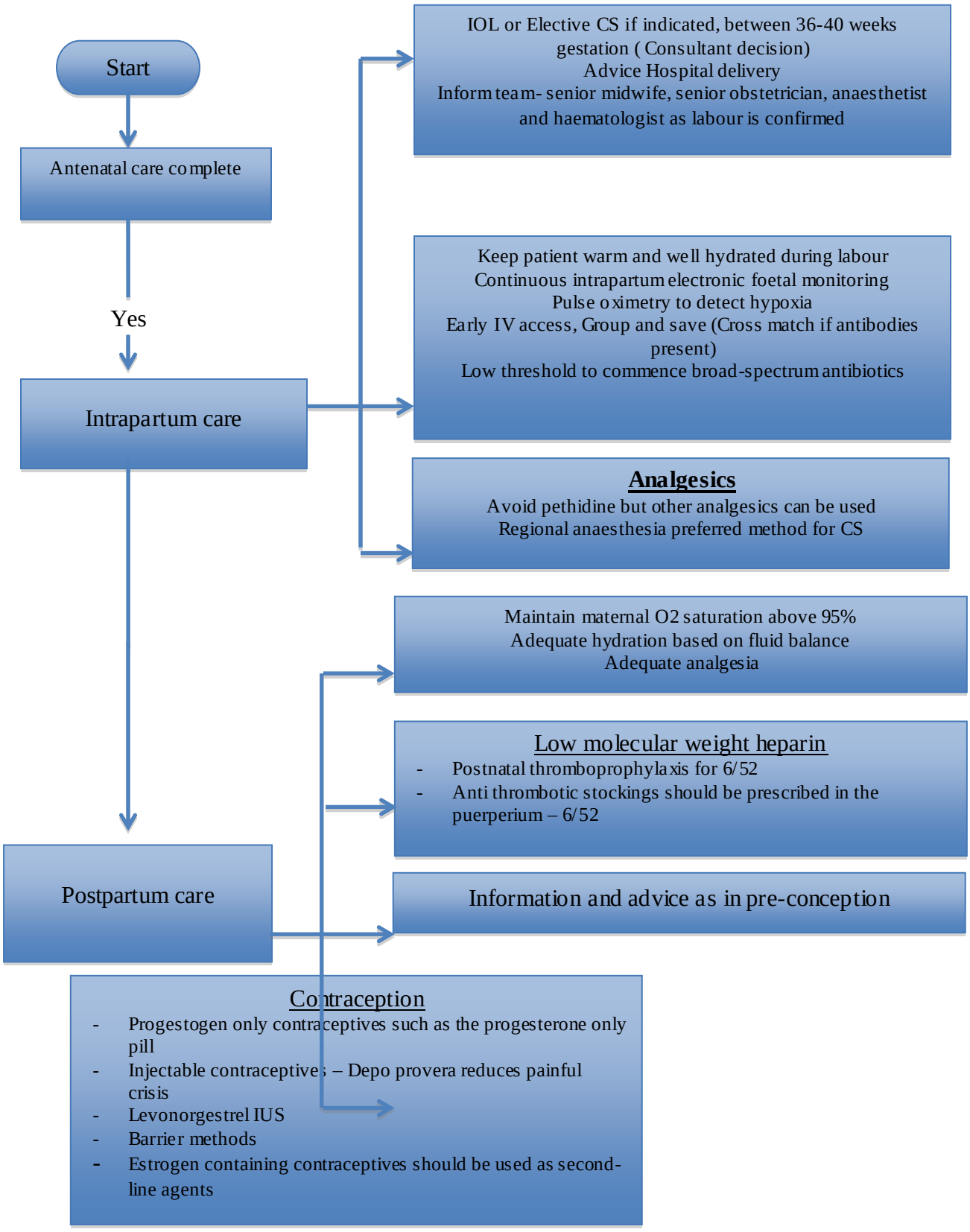
Vaccinations Recommended

- H Influenza B
- Conjugated Meningococcal C
- Pneumococcal
- H1N1
- Hepatitis B

Antenatal Care



Intrapartum and Postnatal Care



Complications

Management of Acute painful crisis in pregnancy

- Rapid clinical assessment
- If pain is severe and oral analgesia is not effective, give strong opioids (e.g. morphine)
- Give adjuvant non-opioid analgesia: paracetamol, NSAID (if 12–28 weeks of gestation)
- Prescribe laxatives, antipruritic and antiemetic if required
- Monitor pain, sedation, vital signs, respiratory rate and oxygen saturation every 20–30 minutes until pain is controlled and signs are stable, then monitor every 2 hours (hourly if receiving parenteral opiates)
- Give a rescue doses of analgesia if required
- If respiratory rate is less than 10/minute, omit maintenance analgesia; consider naloxone
- Consider reducing analgesia after 2–3 days and replacing injections with equivalent dose of oral analgesia
- Discharge the woman when pain is controlled and improving without analgesia or on acceptable doses of oral analgesia
- Arrange any necessary home care and outpatient follow-up appointment

Management of Acute Chest Syndrome(ACS)

- ACS is the second most common complication, reported in 7–20% of pregnancies.
- Tachypnoea, chest pain, cough and shortness of breath in the presence of a new infiltrate on the chest X-ray
- Intravenous antibiotics, oxygen
- Involve Haematologist and Critical care team
- Blood transfusion – Top up if Hb< 6.5g/dl or Exchange transfusion in severe hypoxia with normal Hb.
- Thromboprophylaxis

Auditable standards

- Monitor all patients who are carriers and with Sickle cell disease.
- Appropriate screening for sickle cell disease, haemoglobinopathies
- Appropriate referral pathways
- Notification to the risk management team
- Appropriate training of the obstetric team (midwifery and medical staff).

Appendix 4

References

Sickle cell disease in pregnancy and management – RCOG green top guidelines-61

Appendix 5

This policy has undergone an equality impact assessment screening process using the toolkit designed by the NHS Centre Equality & Human Rights. Details of the screening process for this policy are available from the policy owner.