Urgent Clinical Commissioning Policy Statement: Prenatal surgery for open spina bifida

NHS England Reference: 170049P

1 Equality statement

Promoting equality and addressing health inequalities are at the heart of NHS England's values. Throughout the development of this policy statement, we have:

- given due regard to the need to eliminate discrimination, harassment and victimisation, to advance equality of opportunity, and to foster good relations between people who share a relevant protected characteristic (as cited under the Equality Act 2010) and those who do not share it; and
- given regard to the need to reduce inequalities between patients in access to, and outcomes from healthcare services and to ensure services are provided in an integrated way where this might reduce health inequalities.

In the interests of delivering an urgent commissioning position, a rapid initial equality impact assessment has been carried out.

2 Background

Approximately 700 women per year in the UK will have an ultrasound scan performed in the first half of pregnancy that will diagnose the fetus as having a major form of open spina bifida (myelomeningocoele or myeloschisis).

Babies born with this condition are often unable to walk, incontinent of urine and faeces, may develop hydrocephalus due to incomplete closure of the spinal canal and require a series of operations to drain fluid from the brain later in life (shunt placement). For this reason many parents in the UK opt for a termination of pregnancy shortly after diagnosis although some choose to continue with the pregnancy.

The current UK management would be to close the defect over the spinal cord immediately after birth (neonatal surgery) to reduce the hydrocephalus. However, by birth the damage to the spinal cord has already occurred.

A large multicentre randomised controlled trial in the USA (MOMS) has shown that it is feasible to operate on the fetus before 26 weeks gestation, whilst it is still in the maternal uterus, and to close the defect. The surgery involves opening the uterus as for a caesarean section, exposing the open spina bifida without delivering the fetus, closing the defect and then repairing the uterus.

The benefits to the fetus are that, compared to postnatal treatment, prenatal closure has been shown to improve neurological function and reduce the need for shunt placement. The benefits need to be weighed against the known increase in preterm birth, uterine dehiscence and potential for morbidity in future pregnancies in women who undergo fetal surgery, meaning that counselling of mothers and their partners is particularly important.

Prenatal closure to prevent the hydrocephalus associated with this condition

could be offered as an alternative to postnatal closure.

Data from current European Open Fetal Surgery Centres suggest that only 20% of women with an affected fetus for whom this is an option (i.e. they have chosen not to have a termination of pregnancy) will choose to undergo fetal surgery treatment meaning that they might operate on around 10-20 cases per year in the UK.

3 Evidence Summary

NHS England has considered the evidence submitted as part of the preliminary policy proposal. This has been assessed as requiring an urgent clinical commissioning policy statement. This statement includes the clinical criteria for initiating and for discontinuing this treatment. The statement is informed by up to three of the most clinically considered important publications, identified using a literature search strategy defined by the clinical lead. These publications are summarised below.

Publication 1 (Tulipan et al 2015)

Tulipan et al (2015) reported the Management of Myelo-meningocoele study (MOMS). 91 women were randomised to prenatal surgery and 92 to postnatal repair. The primary outcome was a composite of fetal loss or any of the following: infant death, cerebrospinal fluid (CSF) shunt placement or meeting pre specified criteria for shunt placement. The primary outcome occurred in 73% of patients in the prenatal surgery group and 98% in the postnatal group (P< 0.0001). Actual rates of shunt placement were 44% and 84% in the two groups respectively.

Publication 2 (Adzick et al 2011)

An earlier publication on the same MOMS study reported outcomes on 158 evaluable patients at 30 months. Prenatal surgery resulted in improvement in the composite score for mental development and motor function at 30 months (P = 0.007) and in improvement in several secondary outcomes, including hindbrain herniation by 12 months and ambulation by 30 months.

However, prenatal surgery was associated with an increased risk of preterm delivery and uterine dehiscence at delivery.

4 Commissioning Position

Rationale for a clinical commissioning policy statement

Prenatal surgery to treat open spina bifida is considered to be of such significant clinical importance that an immediate clinical commissioning policy statement has been adopted. The time taken to develop a full clinical commissioning policy proposition for relative prioritisation and implementation would not meet the immediate need for patients, clinicians and the NHS to have clarity about whether an intervention is or is not routinely commissioned.

An urgent commissioning position is appropriate to enable rapid commissioning on the basis that there are good results from recent randomised trials in a small number of patients. These studies suggest that prenatal is effective for patients affected by open spina bifida.

Clinical commissioning position

Based on a limited scoping of the evidence, NHS England has concluded that there is sufficient evidence to support the routine commissioning of prenatal surgery for open spina bifida meeting the clinical criteria listed.

Clinical commissioning criteria

Pre-natal surgery for open spina bifida is commissioned for patients who meet the following criteria:

Fetal congenital open spina bifida at level T1 (first thoracic vertebra) through S1 (sacral segment) with hindbrain herniation.

Inclusion criteria (as per MOMS protocol):

- Gestational age of 19+0 to 25+6 weeks gestation
- Known maternal HIV, HBV, HBC status for inclusion in the management plan.
- No serious maternal medical complications
- Normal karyotype
- Ultrasound confirms congenital open spina bifida below T6;
- Fetal MRI confirms Arnold Chiari Malformation;
- Amniocentesis (to provide a genetic diagnosis to rule out syndromic congenital abnormality which would contraindicate surgery e.g. trisomy 18);
- Fetal lateral ventricles below 20mm, (subject to clarification regarding level of benefit being reduced where there is ventriculomegaly >15mm);
- No additional structural or functional fetal anomalies.

Exclusion Criteria (as per MOMS protocol):

- Multiple pregnancy
- Other fetal anomaly not related to open spina bifida which is likely to significantly impact on fetal surgery or the short or long term outcome;
- Ultrasound confirms fetal spinal kyphosis of 30 degrees or more

- Current or planned cervical cerclage (a stitch placed around the cervix to keep it closed during pregnancy or documented history of cervical insufficiency.
- Obstetric complications such as placenta previa or previous placental abruption
- Short cervical length < 20 mm measured by transvaginal ultrasound
- Obesity as defined by body mass index of 40 or greater
- Previous spontaneous singleton delivery prior to 37 weeks as a contra-indication to a safe near term delivery
- Maternal-fetal Rh isoimmunisation, Kell sensitisation or a history of neonatal alloimmune thrombocytopenia
- Uterine anomaly such as large or multiple fibroids or Mullerian duct abnormality
- Other maternal medical condition which is a contraindication to surgery or general anaesthesia.
- A previous hysterotomy in the active segment of the uterus (whether from a previous classical caesarean, uterine anomaly such as an arcuate or bicornuate uterus, major myomectomy resection, or previous fetal surgery). A previous uncomplicated caesarean section scar is acceptable.
- Inability to comply with the travel and follow-up requirements needed by the Open Fetal Surgery centre
- Maternal hypertension which would increase the risk of preeclampsia or preterm delivery (including, but not limited to: uncontrolled hypertension, chronic hypertension with end organ damage and new onset hypertension in pregnancy).

The following data collection will be required:

i) Fetal or neonatal death;
ii) Placement of CSF shunt by 12 months;
iii) Motor and developmental outcomes at 30 months;
iv) Maternal complications (such as uterine dehiscence);
v) Follow up status.

<u>Clinical commissioning policy development plan</u>

It has been assessed that the development of a full policy is not needed at this time. The present urgent policy statement will be reviewed when new evidence becomes available that may suggest a significant change in eligibility criteria may be indicated.

Should the subsequent published commissioning policy be revised to 'not routinely commissioned', patients started on treatment under this policy statement will continue to have access to it provided they and the clinician responsible for their care continue to believe that it is the right treatment for them.

5 Mechanism for funding

NHS England will reimburse activity undertaken within the terms of this policy statement, as follows: the treatment will only be provided at centres with proven expertise in prenatal surgery for open spina bifida.

6 Date of policy statement approval and review

The policy statement is effective from April 2018.

A clinical commissioning policy is not planned to be developed at this stage. If a clinician, supported by peers, seeks a reappraisal by the Clinical Panel then a new 'Preliminary Policy Proposition' should be submitted.

For guidance email Edmund.jessop@nhs.net.

This policy statement will be formally reviewed when new research evidence becomes available that may suggest a significant change in eligibility criteria may be indicated.

7 References

Adzick NS et al 2011. A randomised trial of prenatal versus postnatal repair 0f myelomeningocoele. New Eng J Med 364: 993 – 1004.

Tulipan N et al. 2015. Prenatal surgery for myelomeningocoele and the need for cerebrospinal fluid shunt placement. J Neurosurg Pediatr 16:613 – 620.