

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No:	1782
Service	Open Fetal Surgery to treat fetuses with 'congenital open spina bifida', (the main types of which are myelomeningocele and myeloschisis)
Commissioner Lead	<i>Bernie Stocks</i>
Provider Lead	<i>Subject to process</i>

1. Scope

1.1 Prescribed Specialised Service

This service specification covers the provision of open fetal surgery and supporting medical services by Fetal Surgery Centres (FSC's) for congenital open spina bifida, the main forms of which are myelomeningocele and myeloschisis. The service will be delivered in a shared care pathway together with existing Regional Fetal Medicine centres (RFMUs) and existing regional neurosurgery centres. This does not include fetoscopic surgery for which the evidence base is currently not sufficient.

1.2 Description

Assessment, open fetal surgery and supporting medical services for women carrying a Fetus with congenital open spina bifida.

1.3 How the Service is Differentiated from Services Falling within the Responsibilities of Other Commissioners

The service is accessible to all pregnant women with a fetus which has a confirmed diagnosis of 'congenital open spina bifida'. All eligible women will have access to care and treatment irrespective of their sexual orientation, gender, race, disability, psycho-social circumstances or geographical location. An important feature of all services is that appropriate pathways are developed for socially disadvantaged and vulnerable people who are often difficult to engage. The specification will cover the cost of activity undertaken by providers commissioned to provide this service, but not any activity undertaken locally by other providers as part of a shared care arrangement.

2. Care Pathway and Clinical Dependencies

2.1 Care Pathway

The service is to provide prenatal surgery for women with a fetus affected by open congenital spina bifida, which meet the inclusion and exclusion criteria to ensure

that there is clear evidence of benefit to the fetus compared with the alternative management of postnatal surgery. Local maternity units will refer women with a fetus suspected of having spina bifida to a RFMU which will then assess and refer onward to one of the FSC's. It is expected that only some ten to 20 women per year will have a fetus with the significant level of spina bifida which meets the evidence-based inclusion criteria.

The service will be provided by a small number (up to two) FSC's which will provide assessment, diagnosis, counselling and open pre-natal surgery for congenital open spina bifida by multi-disciplinary teams (MDT) experienced in the management of this condition. FSC teams will invite the relevant RFMU and neurosurgeons to join discussions and, following diagnosis and the setting out of a treatment plan, there will be shared care with the relevant RFMU. The implementation and delivery of this specification by FSC's will include the development of clear written guidance and shared care agreements. Women should be informed that they should have a planned Caesarean section around 37 weeks for all subsequent pregnancies to avoid rupture of uterus during labour due to the scar on the uterus from the open fetal surgery procedure.

Each FSC will: i) Have an expert MDT, (see personnel list below). This will be led by a Fetal Medicine specialist (this should be an obstetrician who is trained to undertake uterine surgery and has expertise in open fetal surgery for spina bifida repair; ii) Have an appropriate caseload to maintain expertise in the treatment of the condition; iii) Provide a high quality expert service which will: review and co-ordinate the delivery of care with referring RFMU; ensure that referring staff in local maternity units and RFMU's are aware of the proposed care plan for diagnosis and treatment by the FSC; the post-surgical support and management needs of the patient in the remaining gestational weeks iv) Provide robust patient support services.

Follow up requirements for the patient following in utero surgery until the baby is born: The mother will need to attend the RFMU for regular post-surgical follow up assessment as stipulated in the pathways set out in this document to ensure the ongoing safety of the pregnancy.

Components of the FSC MDT:

- a) **Assessment MDT to agree on suitability for surgery:** Core team - fetal medicine specialist, obstetrician who is a fetal medicine specialist with training in uterine surgery undertaking open fetal surgery for spina bifida repair; fetal medicine midwife, neurosurgeon, counsellor, fetal neuroradiologist to review MRI.
- b) **Management MDT to plan the surgery:** All of the above staff plus neonatologist, obstetric anaesthetist with expertise in fetal anaesthesia; theatre team staff for equipment planning and instruments.
- c) **Theatre team for surgery:** Two surgeons with expertise in open fetal surgery for spina bifida repair; two paediatric neurosurgeons/paediatric surgeons; two fetal medicine specialists; (not all need to be in theatre at the same time); two obstetric anaesthetists with expertise in fetal anaesthesia; two operating department practitioners; two obstetric theatre nurses; one neurosurgical scrub nurse; one neonatal consultant; neonatal resuscitation team.
- d) **Recovery staff:** Post-operative recovery staff.

Inclusion criteria/ Mother:

- Singleton pregnancy;
- No serious maternal medical complications;
- Maternal cervix long and closed;
- Known maternal HIV, HBV, HBC status for inclusion in the management plan.

Inclusion criteria/Fetus:

- Ultrasound confirms fetal congenital open spina bifida located somewhere between levels T1 (first thoracic vertebra) through to S1 (sacral segment) with hindbrain herniation.
- Gestational age of 19+0 to 25+6 weeks gestation;
- Amniocentesis or chorionic villus sampling with karyotype or microarray (to provide a genetic diagnosis to rule out; syndromic congenital abnormality which would contraindicate surgery e.g. trisomy 18);
- Fetal kyphoscoliosis less than 30 degrees;
- Fetal MRI confirms Arnold-Chiari II Malformation.

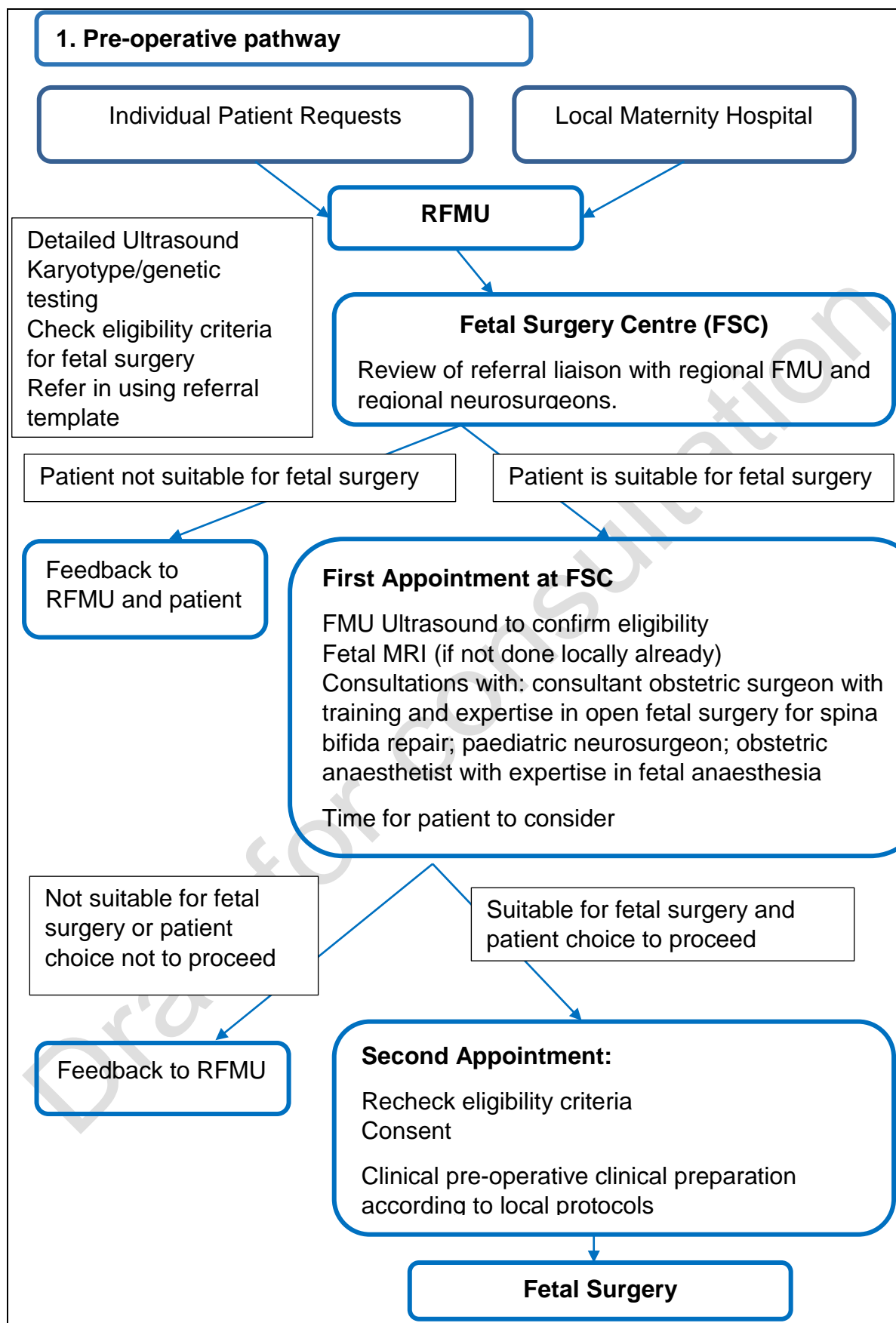
Exclusion Criteria/Mother:

- Multifetal pregnancy;
- Previous spontaneous singleton delivery prior to 37 weeks as a contra-indication to a safe near term delivery;
- Current or planned cervical cerclage (a stitch placed around the cervix to keep it closed during pregnancy) or documented history of cervical insufficiency;
- 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;
- Maternal-fetal Rhesus isoimmunisation, Kell sensitisation or a history of neonatal alloimmune thrombocytopenia;
- Uterine anomaly such as large or multiple fibroids or Mullerian duct abnormality;
- A previous hysterotomy in the active segment of the uterus (whether from a previous classical caesarean, uterine anomaly such as arcuate or bicornuate uterus, major myomectomy resection, or previous fetal surgery) A previous uncomplicated caesarean section scar is acceptable;
- Other maternal medical condition which is a contraindication to surgery or general anaesthesia;
- 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).

Exclusion criteria /Fetus:

- Other fetal anomaly not related to open spina bifida which the MDT review considers is likely to significantly impact on fetal surgery or the short or long term outcome for the baby.

Care pathway: See the flowcharts below for: 1. Pre-operative pathway; 2. Peri-operative Pathway; 3. Post-operative Pathway (Inpatient); 4. Post-operative Pathway (Outpatient); 5. Postnatal Pathway (Neonatal).



2. Peri-operative Pathway

Day before surgery: Patient

- Arrive at Fetal Surgery Centre
- Check consent & eligibility
- Stay at accommodation arranged by hospital or in hospital
- Ultrasound for fetal viability/position and cervical length
- Routine pre-operative preparation

Day before surgery: FSC

- Team Meeting
- Ensure preparations complete as per local protocol
- Neonatal review including to ensure plans of resuscitation of the fetus have been agreed with the patient and documented
- Confirm consent with patient

Day of surgery:

Pre-operative preparation according to agreed local protocols.

Review by: appropriate team members

Bedside ultrasound scan for fetal viability and position

Re-confirm consent

Surgery

Post-operative:

Return to High Dependency Unit/Close Observation Bay

3. Post-operative Pathway (Inpatient)

Day 0: Close Observation Bay / High Dependency Unit

- Routine postoperative recovery and observations
- Post-operative review by surgical and anaesthetic teams
- Bedside Ultrasound for viability
- Immediate post-operative care as per local protocols. Consider tocolysis antibiotics, low molecular weight heparin and analgesia

Day 1 and 2: Close Observation Bay/ High Dependency Unit

- Surgical and anaesthetic review; Ultrasound as per local protocol e.g for viability, membrane separation, cervical length, ductus arteriosus if using indometacin as tocolysis
- Remove catheter once epidural out
- When patient well, consider transfer to antenatal ward

Days 3-7: Antenatal ward

- Modified bed rest (can mobilise to toilet)
 - Ultrasound scan as per local protocol; for fetal viability whilst inpatient ; full FMU ultrasound scan prior to discharge
 - Consider MRI prior to discharge
- Aim to discharge home if patient well and condition stable, otherwise stay until well or transfer to RFMU, considering distance from home.

4. Post-operative Pathway (Outpatient)

Discharge home:

Weekly (or two-weekly if patient's condition is stable) RFMU Ultrasound and review:

- Ultrasound for amniotic fluid volume, Doppler scan, cervical length
- Review for symptoms of leaking fluid or abdominal pain

4-Weekly RFMU detailed review:

- Growth scan, amniotic fluid, Doppler scans, full neural Ultrasound
- One further fetal MRI during pregnancy, ideally 30-36/40 weeks gestation

If any concerning signs or symptoms, discuss with fetal surgery team: Contact - in hours and out of hours – as per individual provider

Spontaneous rupture of the membranes (SROM)/ oligohydramnios/ membrane separation/ cervical shortening
Arrange admission to RFMU and discuss with FSC.

Confirmed preterm labour or fetal distress:

Aim for delivery by emergency Caesarean section (increased risk of uterine rupture). Aim for delivery in RFMU with tertiary level NICU facilities.

Planned RFMU Delivery (with onsite NICU and existing pathway for antenatal paediatric neurosurgical review of babies born with neural tube defects).

5. Postnatal Pathway (Neonatal)

Immediate:

Admission to regional neonatal unit

Day 0:

- Wound review and photograph (with parental consent)
- Neurosurgery, neurology and urology reviews
- Indwelling urinary catheter
- Consider transfer to neurosurgery unit if neurosurgical intervention required (e.g. shunt placement, wound repair, Arnold-Chiari II decompression).

Inpatient:

- Daily head circumference measurement (occipito frontal circumference)
- Renal, hip and head Ultrasound
- Consider MCUG, antibiotics at discretion of urology team
- Orthopaedic review

Discharge from hospital

Usual follow up in regional spina bifida clinic

Includes six week and three month follow up

Description of Pathway Steps	
Steps	Sub-steps
A) Initial diagnosis (local hospital);	Suspected isolated congenital open spina bifida at fetal anomaly ultrasound scan which satisfies the above inclusion and exclusion criteria.
B) Local maternity unit referral to Regional Fetal Medicine Unit (RFMU) after identification of congenital open spina bifida;	Review within 5 days for initial counselling by local fetal medicine specialist and make onward referral to RFMU for confirmation of diagnosis and to discuss options.
C) Referral to SHINE patient support charity;	For support and education for women and their partners/families. https://www.shinecharity.org.uk/ 01733 555988
D) RFMU – assessment;	<ol style="list-style-type: none"> 1. Comprehensive anatomical ultrasound to include documentation of estimated fetal weight, gestational age biometry, lesion level, ventricular size, foot positioning, and lower extremity movement. 2. Ultrasound confirmation that fetus has an isolated congenital open spina bifida which satisfies the above inclusion and exclusion criteria for open fetal surgery. (n.b the RFMU specialist will undertake a detailed ultrasound regardless of whether the woman decides to go ahead with fetal surgery). This includes detailed examination of the fetal heart. 3. Assessment that woman fulfils the maternal inclusion and exclusion criteria for open fetal surgery.
E) RFMU parental education;	<ol style="list-style-type: none"> 1. RFMU undertakes education of the woman (and family) about neural tube defects, community resources, provides initial information regarding prenatal and postnatal surgery. 2. Provides Patient Information Leaflet: Fetal Spina Bifida and Surgical Closure during Pregnancy, provides information on management following prenatal surgery, and recommendations for continued care in the postnatal period.

F) RFMU counselling on options for fetus;	<ol style="list-style-type: none"> 1. RFMU to arrange counselling by regional paediatric neurosurgeon if available, neonatologist; fetal medicine midwife; and maternal-fetal medicine specialist. 2. Counselling to include: <ul style="list-style-type: none"> • discussion of termination of pregnancy (ToP) as part of informed choice; • surgery – including that there are pre and post-natal surgery options; provide patient information leaflet about fetal surgery • Risk of aneuploidy – offer invasive prenatal diagnosis (generally an amniocentesis then karyotype or microarray depending on local policy, chorionic villus sampling also an option if gestational age for amniocentesis not reached yet).
G) Further RFMU investigations if diagnosis of congenital open spina bifida is confirmed (MRI, amnioscentesis);	<ol style="list-style-type: none"> 1. If diagnosis confirmed and woman consents, then MRI and amniocentesis plus karyotype or microarray. 2. If above criteria met, complete referral to Fetal Surgery Centre (FSC) if the woman is interested.
H) RFMU referral to Fetal Surgery Centre (FSC);	<ol style="list-style-type: none"> 1. If woman wishes to consider pre-natal surgery, RFMU to contact FSC receiving unit. 2. Referral made to receiving FSC on standardised proforma for anonymised information and ultrasound and MRI images for review by receiving surgeon with expertise in open fetal surgery for spina bifida repair; (pre-transfer selection tool). 3. Teleconference with the woman, referring RFMU and receiving FSC if appropriate to discuss the case, (or outpatient consultation if appropriate). 4. If receiving FSC accepts, proceed to complete referral paperwork.
<p>I) FSC Assessment to confirm fetal and maternal condition suitable for open fetal surgery.</p> <p>And FSC detailed current counselling specific to prenatal surgery for congenital open spina bifida;</p>	<p>On the same day, the following will happen:</p> <ol style="list-style-type: none"> 1. Comprehensive anatomical fetal ultrasound to confirm congenital open spina bifida diagnosis with gestational age biometry, lesion level, ventricular size, foot positioning, and lower extremity movement. 2. Fetal echocardiogram. 3. Fetal MRI undertaken if not already performed or if previous MRI at RFMU not sufficient) to document hindbrain herniation and exclude anomalies not associated with spina bifida that have not been detected by ultrasound examination. 4. Review of fetal MRI by neuroradiologist. 5. Review of maternal history, fetal karyotypic findings 6. If the findings above still satisfy the inclusion and exclusion criteria for open fetal surgery, counselling of woman in detail again by obstetric surgeon who is a Fetal Medicine specialist who is trained to undertake uterine surgery and has expertise in open fetal surgery for spina bifida repair;

	<p>neonatologist, neurosurgeon, obstetric anaesthetist with expertise in fetal anaesthesia (including undertaking pre-operative assessment and fetal medicine midwife.</p> <ol style="list-style-type: none"> 7. Explain risks including 'low' risk of (fetal) demise and extreme premature delivery, plus high dependency unit environment post-operatively, although anaesthetic risk to patient is as from any surgery. 8. Explain the current outcomes evidence for prenatal surgery and local data 9. Explain data regarding open form of prenatal surgery overwhelmingly outweighs limited data from fetoscopic surgery currently. 10. Explain steps during fetal surgery, what will happen to mother during surgery and aftercare. 11. Explain need for all further deliveries to be by Caesarean section. 12. Woman asked to consider all information provided, to discuss with her family and local RFMU if needed and let FSC know her decision about fetal surgery, postnatal surgery or ToP.
J) FSC – pre surgical follow up by telephone or in person depending on logistical considerations	<ol style="list-style-type: none"> 1. Woman has sufficient time to discuss with family and local RFMU and/or local obstetric team about fetal surgery option. 2. Woman signs consent form for surgery. Decision with woman, fetal medicine consultant and neonatologist about fetal resuscitation and/or deliver if an emergency arises during the fetal surgery.
K) Fetal surgery procedure	<ol style="list-style-type: none"> 1. The day before surgery a repeat ultrasound will be performed to get accurate Estimated Fetal Weight, and to do a final confirmation of suitability for open fetal surgery (no new findings that contraindicate benefit). 2. Drugs for fetal resuscitation made up by Pharmacy prepared in advance according to the Estimated Fetal Weight. 3. The woman will be fasted overnight and surgery will be performed in theatre. 4. Under general anaesthetic and epidural anaesthesia, a maternal laparotomy and a hysterotomy will be performed under ultrasound guidance to determine the optimum incision. The fetus will receive anaesthesia by intramuscular injection. The spinal defect will be positioned in the hysterotomy and will be closed surgically by a Paediatric Neurosurgeon with experience of postnatal and prenatal spina bifida closure. The fetal heart will be monitored by ultrasound by a Fetal Medicine specialist with expertise of fetal echocardiography, throughout with recourse to fetal resuscitation if necessary. The uterus and abdomen will be closed 5. The woman will remain in the FSC high dependency unit for 24 to 48 hours before returning to the FSC postnatal ward,

	<p>as clinically required.</p> <ol style="list-style-type: none"> 6. Ultrasound surveillance of uterine scar for bleeding and amniotic fluid leakage. 7. Post-surgery, the FSC will contact the referring RFMU and arrange the date of transfer of care of the management of the woman back to the RFMU. 8. FSC to provide management advice to regional NHS referring centre neurosurgery department re post-delivery care. 9. Woman goes home.
L) RFMU follow up during remainder of pregnancy	<ol style="list-style-type: none"> 1. RFMU manages care including follow up ultrasound scans and MRI according to FSC protocol to include reversal or Arnold-Chiari II Malformation and surveillance of uterine scar for bleeding and amniotic fluid leakage. 2. Results of follow up ultrasound, MRI scans and maternal condition to be communicated to the FSC directly after imaging. 3. If complications arise, the referring RFMU will communicate with receiving FSC to seek advice.
M) RFMU Delivery;	<ol style="list-style-type: none"> 1. Delivery is by Caesarean section by an experienced obstetric surgeon who will inspect the hysterotomy scar, repair the defect if needed. 2. If the membranes rupture or the woman labours, woman to present to nearest maternity unit along normal neonatal operational delivery network pathway for management of preterm labour, delivery and neonatal critical care and subsequent neurosurgical review. 3. Individualised care plan will be put in place taking into account risk of preterm labour, infection, prematurity and other maternal and fetal factors. 4. If labouring, woman to go to nearest maternity centre, which will then refer her up to the RFMU or call RFMU for telephone advice if time does not permit transfer. 5. Note in clinical notes that all subsequent maternal deliveries will need to be by planned Caesarean Section to avoid uterine rupture in labour.
N) RFMU post-delivery care	RFMU provides detailed information to FSC on outcome of Caesarean section surgery and short term maternal outcome.
O) Regional NHS Neurosurgery and Neonatal/Paediatric Department	<ol style="list-style-type: none"> 1. Regional NHS referring centre neurosurgery and neonatal departments to jointly organise postnatal follow up of the infant, including enhanced developmental support and surveillance. 2. Follow up will be for at least up to two years of age corrected for prematurity as per NICE guidelines for babies born preterm (<37 weeks) if they are considered at high risk of problems and for up to age four for babies born at <28weeks. 3. Communicate outcomes to FSC after each assessment.

P) FSC follow up	FSC arranges local follow up of woman (mother) and neonate for review by the fetal surgery team and neurosurgeons.
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2.2 Interdependence with other Services

Clinical co-dependencies include:

- **Local Maternity Unit:** Obstetrics and Paediatrics/Neonatology
- **Regional neurosurgery units**
- **Regional Fetal Maternity Unit (RFMU)** (may be onsite at District General Hospital or referred across): Obstetrics; Fetal Medicine; Neonatal medicine (NICU); Paediatric neurosurgery; Fetal cardiology, Paediatric Neurology/Neurodisability, Urology, Paediatric Radiology. Pathways to paediatric neurosurgery.
- **National Fetal Surgery Centre (FSC): each FSC will have the following in place:**
 - a) **Infrastructure** - Co-located: fetal surgery clinic capacity, fetal MRI neuroradiology expertise, neonatal intensive care unit (NICU), obstetric high dependency unit, adult intensive care unit, inpatient ward for the care of pregnant women which is staffed by midwives to monitor fetus and nurses, fetal medicine theatre lists, theatres.
 - b) **Equipment:** technology for MDTs
 - c) **Staffing:** Obstetrics; Fetal medicine; Fetal Surgery, Neonatal medicine (NICU); Paediatric neurosurgery; Paediatric neurodisability; obstetric anaesthetist with expertise in fetal anaesthesia, fetal MRI neuroradiology expertise, adult critical care unit.

3. Population Covered and Population Needs

3.1 Population Covered By This Specification

The population covered is all women falling within the direct commissioning responsibilities of NHS England who meet the inclusion and exclusion criteria.

3.2 Population Needs

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 (myelomeningocele 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 women in the UK opt for a ToP shortly after diagnosis, although some choose to continue with the pregnancy.

The benefits to those fetuses which are in scope for pre-natal surgery are that prenatal closure has been shown to improve neurological function, it can reduce the need for shunt placement to cut the risk of hydrocephalus and in some cases, the patient may have improved motor function/be able to walk. 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.

Data from current European Open Fetal Surgery Centres suggest that only 20% of women with an affected Fetus will choose to undergo fetal surgery treatment meaning that they might operate on around 10-20 cases per year from the UK.

3.3 Expected Significant Future Demographic Changes

There has been a gradual decline in the incidence of spina bifida in the UK over the past forty years. In future the incidence is likely to change in line with the birth (pregnancy) rate.

3.4 Evidence Base

This specification has been developed on the basis of clinical consensus, informed by a randomised trial in the USA (MOMS) which has shown that it is feasible to operate on the baby before 26 weeks gestation, whilst it is still in the womb, and to close the defect. The surgery involves opening the uterus as for a caesarean section, exposing the open spina bifida without delivering the baby, closing the defect and then repairing the uterus. (Tulipan et al 2015) and a position statement (Cohen et al 2014). The morbidity of open spinal lesions is dependent primarily on the proximal level of the lesion and the Arnold Chiari 11 Malformation (Ovaere(2014) See Appendix 1 for map of the spine.

- 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.
- Cohen et al (2014) Position Statement on Fetal Myelomeningocele Repair notes that is essential to establish minimum criteria for centres providing open fetal myelomeningocele repair to ensure optimal maternal and fetal/paediatric outcomes, as well as patient safety both short- and long-term; and to advance our knowledge of the role and benefit of fetal surgery in the management of fetal myelomeningocele. [https://www.ajog.org/article/S0002-9378\(13\)00957-5/fulltext](https://www.ajog.org/article/S0002-9378(13)00957-5/fulltext)
- Ovaere et al (2014) notes that the Management of Myelomeningocele Study (MOMS) eventually provided level I evidence that prenatal repair of lesions above S1 between 19 and 26 completed weeks of gestation improved the outcome when compared to postnatal repair. Also that 'Fetal surgery resulted in a decreased need for shunting by 12 months of age (RR: 0.48; 95% CI: 0.36–0.64), improvement in hindbrain herniation and a better composite score for mental development and motor function at 30 months, including a higher rate of ambulation (RR: 2.01; 95% CI: 1.16–3.48).
- Tulipan et al (2015) reported on the Management of Myelo-meningocoele (MOMS) study in which 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.

- Werner et al (2012) Prenatal myelomeningocele repair is cost effective and frequently cost saving compared with post-natal myelomeningocele repair despite the increased likelihood of maternal and future pregnancy complications associated with prenatal surgery...The average gestational age at birth is 3 weeks lower with prenatal surgery (34.1 vs 37.3weeks, P,<0.001) [as compared to postnatal surgery].

4. Outcomes and Applicable Quality Standards

4.1 Quality Statement – Aim of Service

The aim of the service is to reduce the morbidity associated with open spina bifida by providing prenatal corrective surgery to appropriately selected women who meet the inclusion and exclusion criteria.

NHS Outcomes Framework Domains

Domain 1	Preventing people from dying prematurely	✓
Domain 2	Enhancing quality of life for people with long-term conditions	✓
Domain 3	Helping people to recover from episodes of ill-health or following injury	
Domain 4	Ensuring people have a positive experience of care	✓
Domain 5	Treating and caring for people in safe environment and protecting them from avoidable harm	✓

4.2 Indicators Include:

Detailed definitions of indicators, setting out how they will be measured is included in Appendix 3 and Schedule 6 of the NHS England contract

Commissioned providers are required to participate in annual quality assurance and collect and submit data to support the assessment of compliance with the service specification as set out in Schedule 4A-C.

Applicable CQUIN goals are set out in Schedule 4D.

Number	Indicator	Data Source	Outcome Framework Domain	CQC Key question
Clinical Outcomes				
101	Percentage of births within 30 days of prenatal surgery	Provider	1,3,5	safe, effective
102	Percentage of women with uterine rupture	Provider	1,3,5	safe, effective
103	Percentage of neonatal deaths in the last 12 months	Provider	1,3,5	safe, effective
104	Percentage of women treated for post-operative infection	Provider	1,3,5	safe, effective
105	Percentage of mothers with peri-operative pulmonary oedema requiring non-invasive or invasive respiratory support immediately post-surgery	Provider	1,3,5	safe, effective
106	Percentage of in utero deaths following surgery during the last 12 months.	Provider	1,3,5	safe, effective
107	Percentage of fetuses with amniotic fluid index less than fifth centile following surgery	Provider	1,3,5	safe, effective
108	Percentage of fetuses with intra-operative compromise requiring resuscitation	Provider	1,3,5	safe, effective
109	Percentage of fetuses in which a patched closure is required	Provider	1,3,5	safe, effective
110	Percentage of babies with post-natal patch infection	Provider	1,3,5	safe, effective
111	Percentage of babies with post-natal CSF leakage	Provider	1,3,5	safe, effective
112	Percentage of babies with post-natal CSF shunt diversion placements	Provider	1,3,5	safe, effective
113	Percentage of babies having neurosurgical intervention in the neonatal period (up to 28 days of life).	Provider	1,3,5	safe, effective
114	Percentage of babies with recorded assessment of functional motor and sensory level at 30 months	Provider	1,3,5	safe, effective

115	Percentage of babies with recorded complete results of a structured assessment for neurodisability (cognitive and impaired movement) between 24mths to 36months gestationally corrected age.	Provider	1,3,5	safe, effective
116	Percentage of babies with improvement in Arnold-Chiari II malformation	Provider	1,3,5	safe, effective
117	Percentage of babies having surgery for infantile Arnold-Chiari II malformation within 12 months of delivery.	Provider	1,3,5	safe, effective
118	Percentage of women offered an appointment by the Fetal Surgery Centre within one week of referral by the Regional Fetal Medicine Unit	Provider	1,3,5	safe, effective
Patient Experience				
201	There is information for women and families	Self declaration	4	caring, responsive
202	Feedback from women and families is reviewed and informs service development and improvements	Self declaration	4	caring, responsive
203	Women and families receive counselling by the MDT	Self declaration	4	caring, responsive
Structure and Process				
001	There is a specialist multidisciplinary team	Self declaration	1.3.5	safe, effective,
002	There is a competency based training programme in place	Self declaration	1.3.5	safe, effective,
003	There is a multidisciplinary discussion of each referral to agree the diagnosis and treatment plan	Self declaration	1.3.5	safe, effective,
004	There are agreed pathways in place	Self declaration	1.3.5	safe, effective,
005	There are agreed clinical guidelines in place as detailed within the service specification.	Self declaration	1.3.5	safe, effective,
5. Applicable Service Standards				
5.1 Applicable Obligatory National Standards The provider must meet all legal and mandatory requirements for the provision of				

obstetric, paediatric and neurosurgical care

5.2 Other Applicable National Standards to be met by Commissioned Providers

Position statement of fetal myelomeningocele: The provider should meet the standards set out in a position statement published in the American Journal of Obstetrics and Gynaecology: <http://dx.doi.org/10.1016/j.ajog.2013.09.016>

Applicable service standards: NICE guideline on preterm neurodevelopmental follow-up Developmental follow-up of children and young people born preterm' which can be found at: <https://www.nice.org.uk/guidance/NG72>.

5.3 Other Applicable Local Standards

Not applicable

6. Designated Providers (if applicable)

Not applicable.

7. Abbreviation and Acronyms Explained

The following abbreviations and acronyms have been used in this document:

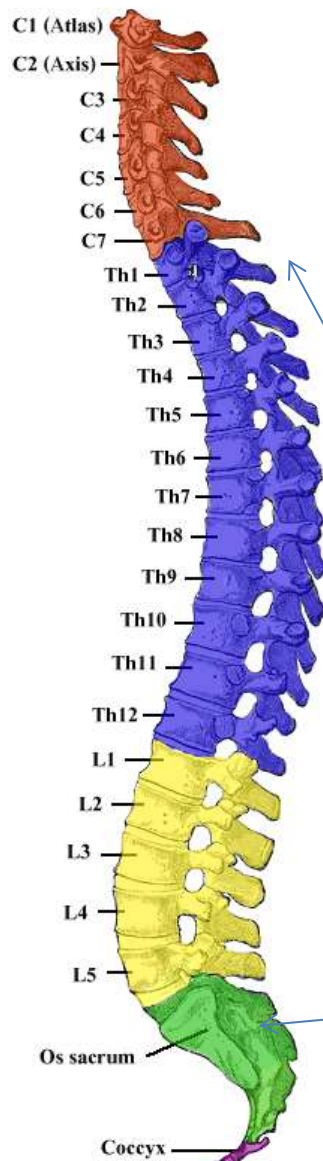
ACM	Arnold-Chiari II Malformation (see also spina bifida aperta below) https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Chiari-Malformation-Fact-Sheet#3
CSF	Cerebrospinal fluid
CRP	C-reactive protein serum concentration
FSC	National Fetal Surgery Centre
HIV	Human Immunodeficiency Virus
HBV	Hepatitis B virus
HBC	Hepatitis C virus
MCUG/ MCU	Micturating cystourethrogram
MDT	Multi-disciplinary team
MRI	Magnetic Resonance Imaging
MOMS	Management of Myelomeningocele clinical trial https://www.ncbi.nlm.nih.gov/pubmed/21306277
NICU	Neonatal Intensive Care Unit
PTL	Preterm Labour
RFMU	Regional Fetal Medicine Unit
S1	First sacral vertebrae (see diagram at Appendix 1)

Spina bifida aperta (open)	The spinal cord and nerve tissue bulge through an open hole or area of thin membrane on the baby's back. Aperta (Latin for 'open'). This form of spina bifida is also called meningocele or myelomeningocele. These defects are often associated with the Arnold-Chiari II malformation.
SROM	Spontaneous rupture of the membranes.
ToP	Termination of pregnancy.
	Ventriculomegaly (also known as hydrocephalus). The brain is surrounded by a clear fluid called cerebral spinal fluid (CSF), although with spina bifida and for other reasons, the parts of the brain called lateral ventricles which are filled with CSF, become enlarged (dilated), which can prevent the correct development of the brain. The most common definition of ventriculomegaly is where the width of the atrium of the lateral ventricle is greater than 10mm.
VP	Ventriculoperitoneal shunt - during shunt surgery, a thin tube called a shunt is implanted in the brain to drain the excess cerebrospinal fluid (CSF) from the brain through the shunt to another part of the body, usually the tummy, where it is then absorbed into the bloodstream.

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Appendix 1: Vertebral Column



The vertebral column is divided into five *regions*, which are - from top to bottom:

- Cervical - Shown in red, this region supports the [head](#). It is made up of 7 vertebrae.
- Thoracic - Shown in blue, this region supports the [ribs](#). It is made up of 12 vertebrae.
- Lumbar - Shown in yellow, this region is located in the lower back. It is made up of 5 vertebrae.
- Sacral - Shown in green. It is made up of 5 vertebrae that are [fused](#) together.
- Coccygeal - Shown in purple. It is made up of 3 to 5 vertebrae

Only spinal lesions that occur anywhere between levels T1 to S1 are in scope for open pre-natal spina bifida surgery.

Appendix 2: Example of the FSC Patient information leaflet for England – including the types of questions and answers that the provider patient information leaflet might include:

- What is spina bifida?
- What does spina bifida mean for my child?: Motor, sensory and orthopaedic impacts; Brain impacts; Bladder and bowel impacts; Hormonal impacts; Psychosocial and sexual impacts
- Should I continue or terminate my pregnancy?
- What if I decide to continue the pregnancy?
- What is fetal surgery?
- What are the benefits of fetal surgery as compared to surgery on the baby after it is born /how can fetal surgery help?
- What are the risks of fetal surgery?
- How should I prepare for fetal surgery?
- Asking for your consent
- What happens during fetal surgery?
- What should I expect after fetal surgery?

What is spina bifida?

Spina bifida arises when in early pregnancy (before the 6th week), the spinal column and spinal cord are not properly formed. It occurs in around 4 in 10,000 pregnancies.

What causes spina bifida?

The exact cause is unknown. Several factors may be responsible such as insufficient folate (a B vitamin) in the diet. There are also hereditary factors.

What does spina bifida mean for my child?: Every person with spina bifida is unique, meaning there is a large variety of possible effects, from mild to severe. Generally, a defect higher up on the back is associated with more serious consequences. Ultrasound of the baby can be used to determine the position (height) of the defect in the spinal column before they are born. This will give doctors information about what problems the child will likely have. Typically these problems will include weakness in the feet or legs and problems with bladder and bowel control. However it is not possible to predict before birth exactly how severe these problems will be. Sometimes other abnormalities are also found during the ultrasound scan. This combination increases the likelihood of an underlying genetic syndrome. A procedure to sample the amniotic fluid (amniocentesis) is recommended to exclude chromosomal problems (chromosomes are the carriers of genetic information inside each of our cells). If additional problems are confirmed, the outcome will depend on all of the abnormalities together. :

- **Motor, sensory and orthopaedic problems:** Normally, nerves from the spinal cord go to the muscles of the legs and feet to allow movement. In spina bifida, the spinal cord and nerves are often entirely or partially interrupted at the place of the lesion in the back. This can cause: deformity of the spine (also called scoliosis), muscle weakness in the legs, impaired balance, contracture of several joints, club feet. Children with a very low lesion (below Sacral 1 (S1)) are likely to be able to stand and learn to walk or have only mild weakness or deformity in the feet. Because nerve

pathways are interrupted in spina bifida, there is less or no sensation in the skin served by nerves below the site of the lesion in the back. This can lead to pressure ulcers in the skin, or it can cause cold feet and lower legs.

- **Brain changes:** The changes in the hindbrain, referred to as Arnold-Chiari II Malformation do not necessarily lead to problems for the child, and will not usually require treatment. In severe cases, children may have difficulty with swallowing and breathing. Brain fluid may continue to accumulate inside the ventricles, leading to ventriculomegaly. This occurs in about half to two thirds of babies with spina bifida. If untreated, hydrocephalus can cause damage to the brain due to increasing pressure. When hydrocephalus occurs, surgeons often need to implant a shunt, a thin tube that drains the fluid from the brain into the abdominal cavity (tummy) of the child. The satisfactory working of the shunt will be assessed from time to time in the outpatient clinic. Further surgery to a shunt may be required if there are problems such as infection or blockage. Such shunt complications can be responsible for long term disabilities. Hydrocephalus and complications of drains may result in some learning and behavioural issues. As many as two thirds of children with spina bifida have an IQ within the normal range. However, children with open spina bifida may have learning and behaviour issues. These include difficulties with concentration, attention and problem solving.
- **Bladder and bowel problems:** Children with spina bifida often have problems controlling their bladder. This may cause urinary incontinence or inability to fully empty their bladder (referred to as urinary retention). This may lead to bladder and/or kidney infections. To prevent complications, medical staff may recommend that they empty their bladder using a small catheter through the urethra several times a day (called clean intermittent catheterisation). First the parents are taught this, later the children can learn to do it for themselves. Children also often have bowel issues, resulting in either incontinence or constipation. This can be helped by bowel wash-outs (irrigation) or medication.
- **Hormonal problems:** Children with spina bifida often have a shorter stature than usual. Early puberty is more common, especially in cases of hydrocephalus.
- **Psychosocial and sexual problems:** When young adults with spina bifida are asked about whether they feel happy, their response usually is very similar to their peers. Men with spina bifida may have decreased fertility, but medication usually helps with erection difficulties. Women with spina bifida may have reduced sensation which affects their full enjoyment of sex.

Should I continue or terminate my pregnancy? Many factors play a role when choosing what to do in your pregnancy. These include the anticipated severity of the condition and how you and your partner and family feel you can cope with caring for a child with this condition.

What if I decide to continue the pregnancy?: If this is your choice then you will be supported throughout your pregnancy. During the last three months of pregnancy, your baby will be monitored by local clinicians ultrasound, for instance to measure the amount of fluid in the brain. If the fluid increases, sometimes the medical staff will recommend that your baby is delivered a little earlier.

What is fetal surgery?: Fetal surgery is where surgeons perform an operation on a baby whilst it is still in the womb, hence before it is born. In a baby with spina bifida, the defect is closed using a similar surgical technique to closure after birth. The womb is then closed and the baby continues to grow and develop in the womb until birth. In 2011, the results of an American study called the "MOMS trial" were published. The study investigated the effect of operating on babies with spina bifida whilst they were still in the womb as compared to operating on babies after birth.

What are the benefits of fetal surgery as compared to surgery on the baby after it is born/How can fetal surgery help? In the MOMS study mentioned above, there were 77 children with spina bifida who were operated on in pregnancy, while they were still in the womb. Their outcomes were compared to 80 babies who had spina bifida surgery after birth. Children who were operated before birth had: i) less severe Arnold-Chiari II Malformation (hindbrain herniation); ii) less need for a shunt for hydrocephalus by the age of one year, though the effect was less when the ventricles were already enlarged before the operation; iii) better leg function at 30 months of age. On further follow-up those operated before birth seemed to have better bladder function, but that evidence is yet not conclusive.

What are the risks of fetal surgery? There are risks of performing the spina bifida closure operation before birth, such as an increased risk of premature labor, premature rupture of membranes and decreased amniotic fluid. Premature delivery (birth before 37 weeks of gestation) can have its own complications that may offset some benefits of the fetal surgery. After an operation during pregnancy, doctors recommend that women deliver their babies by caesarean section, both in the same pregnancy and in future pregnancies. This is to prevent the scar in the womb opening during labour. Women are also asked to wait two years before conceiving again.

How should I prepare for fetal surgery? If you are interested in fetal surgery for your future baby, your Fetal Medicine Consultant will discuss it with you and your partner/family. The doctors will repeat the evaluation of your baby by ultrasound and MRI, before the decision can be made to proceed with surgery. This is to make sure that we know as much as possible about the abnormality that the baby has. The fetal surgery operation should take place before 26 weeks of gestation.

What happens during fetal surgery? You will be asked to come into hospital after fasting from the night before (for morning surgery). You will be shown to a room and given a gown to change into. You will meet the anaesthetist and Fetal Surgery Centre surgical team again. When you are ready you will be taken into the operating theatre and the anaesthetist will offer to insert an epidural catheter (fine plastic tube) into your back for pain relief after the operation. You will then have a general anaesthetic (to put you to sleep). Once you are anaesthetised, a bikini line cut will be made in your abdomen. This is slightly wider than the size of the cut used for Caesarean section birth, but in the same place on your abdomen. The uterus (womb) is then opened to allow access to the baby. The spina bifida defect is examined and closed by the paediatric neurosurgeon. The uterus and then abdomen are both closed. You

will then be woken up.

What should I expect after fetal surgery? When you wake up from the operation you may feel a bit sore, but painkillers will be available for you. You will also have an epidural analgesia catheter in place for pain relief. There will be also be a catheter in your bladder which will be removed once you are able to walk around and pass urine in the toilet. You will stay in hospital for a period of around one week, and when you go home you will be offered regular follow up and advised to rest.

Appendix 3: Complete description of Metrics

Number	Indicator	Descriptor	Notes	Evidence documents	Data Source	O.F Domain	CQC Key question
	Clinical Outcomes - quantitative data						
101	Percentage of births within 30 days of prenatal surgery	Numerator number of births within 30 days of prenatal surgery Denominator number of fetuses having surgery following surgery			Provider	1,3,5	safe, effective
102	Percentage of women with uterine rupture	Numerator number of women with uterine rupture Denominator number of women delivered post surgery			Provider	1,3,5	safe, effective
103	Percentage of neonatal deaths in the last 12 months	Numerator number of neonatal deaths in the past 12 months Denominator number of fetuses having surgery			Provider	1,3,5	safe, effective
104	Percentage of women treated for post operative infection	Numerator number of women treated for post operative infection Denominator Number of women having fetal surgery			Provider	1,3,5	safe, effective

105	Percentage of mothers with peri-operative pulmonary oedema requiring non-invasive or invasive respiratory support immediately post-surgery	Numerator number of women with pulmonary oedema Denominator Number of women having fetal surgery			Provider	1,3,5	safe, effective
106	Percentage of in utero deaths following surgery during the last 12 months.	Numerator number of in-utero deaths in the past 12 months Denominator number of fetuses having surgery			Provider	1,3,5	safe, effective
107	Percentage of fetuses with amniotic fluid index less than fifth centile following surgery	Numerator number of fetuses with amniotic fluid index less than fifth centile following surgery Denominator number of fetuses having surgery			Provider	1,3,5	safe, effective
108	Percentage of fetuses with intra-operative compromise requiring resuscitation	Numerator number of fetuses with intra-operative compromise requiring resuscitation Denominator number of fetuses having surgery			Provider	1,3,5	safe, effective
109	Percentage of fetuses in which a patched closure is required	Numerator number of fetuses having had prenatal surgery with patch closure of MMC Denominator			Provider	1,3,5	safe, effective

		number of fetuses having surgery					
110	Percentage of babies with post natal patch infection	Numerator number of babies with post natal infection of patch Denominator Number of fetuses having had patch surgery			Provider	1,3,5	safe, effective
111	Percentage of babies with post natal CSF leakage	Numerator number of babies having post natal CSF leakage Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
112	Percentage of babies with post natal CSF shunt diversion placements	Numerator number of babies having post natal CSF shunt diversion placements Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
113	Percentage of babies having neurosurgical intervention in the neonatal period (up to 28 days of life)	Numerator number of babies having post natal surgery Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
114	Percentage of babies with recorded assessment of functional motor and sensory level at 30months	Numerator number of babies with recorded assessment of functional motor and sensory level at 30 months Denominator			Provider	1,3,5	safe, effective

		number of births following fetal surgery					
115	Percentage of babies with recorded complete results of a structured assessment for neurodisability (cognitive and impaired movement) between 24mths to 36months gestationally corrected age.	Numerator number of babies with recorded complete results of a structured assessment for neurodisability (cognitive and impaired movement) between 24mths to 36months gestationally corrected age. Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
116	Percentage of babies with improvement in Arnold-Chiari II malformation	Numerator number of babies with improvement in Arnold-Chiari II malformation Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
117	Percentage of babies having surgery for infantile Arnold-Chiari II malformation within 12 months of delivery.	Numerator number of babies having surgery for Arnold-Chiari II malformation within 12 months of delivery Denominator number of births following fetal surgery			Provider	1,3,5	safe, effective
118	Percentage of women offered an appointment by the Fetal Surgery Centre	Numerator number of women offered an appointment by the Fetal Surgery Centre within one			Provider	1,3,5	safe, effective

	within one week of referral by the Regional Fetal Medicine Unit	week of referral by the Regional Fetal Medicine Unit Denominator number of women referred					
Patient Experience							
201	There is information for women and families	<p>The service should have written information for women and families covering at least the following:</p> <ul style="list-style-type: none"> - information about the service including names and functions/roles of the specialist team - description of the pathway <p>The information should have been approved</p> <ul style="list-style-type: none"> - information about the services offering psychological, social and spiritual/cultural support including SHINE support group. - information about treatment including possible interventions , benefits and side effects - relevant contact points. 	<p>The information should be agreed through the Trusts governance processes and it is recommended that the information is available in languages and formats understandable by patients and carers including local ethnic minorities and people with disabilities.</p>	Operational Policy.	Self declaration	4	caring, responsive
202	Feedback from women and families is reviewed and informs service development and improvements	The service should have undertaken an exercise to obtain feedback on the women's and families' experience of the services		Annual report	Self declaration	4	caring, responsive

		<p>offered.</p> <p>The exercise and results should have been presented to and discussed at divisional or directorate level.</p>					
203	Women and families receive counselling by the MDT	<p>Women and families must be given counselling by members of the MDT including including obstetric surgeon with training in fetal surgery and expertise in open fetal surgery for spina bifida repair, neonatologist, neurosurgeon, obstetric anaesthetist and fetal medicine midwife prior to surgery.</p>		Operational policy	Self declaration	4	caring, responsive
Structure and Process - infrastructure requirements, staffing, facilities etc							
001	There is a specialist multidisciplinary team	<p>There is a specialist multidisciplinary team which includes:</p> <ul style="list-style-type: none"> - 2 fetal medicine specialists - obstetric surgeon with training in fetal surgery and expertise in open fetal surgery for spina bifida repair - fetal medicine midwife - paediatric neurosurgeon - neonatologist - fetal neuroradiologist - 2 obstetric anaesthetists with expertise in fetal anaesthesia 		Operational Policy	Self declaration	1.3.5	safe, effective,

		<ul style="list-style-type: none"> - obstetrician - psychologist / counsellor <p>all above should have experience in fetal surgery and have this specified within their job plans/timetable</p>					
002	There is a competency based training programme in place	There must be a competency based training programme in place for health care professionals involved in the care of women having fetal surgery		Training programme	Self declaration	1.3.5	safe, effective,
003	There is a multidisciplinary discussion of each referral to agree the diagnosis and treatment plan	<p>There is a multidisciplinary discussion for agreeing the diagnosis and treatment plan of women referred for fetal surgery. The discussion should include at least the following:</p> <ul style="list-style-type: none"> - fetal medicine specialist - obstetric surgeon with training in fetal surgery and expertise in open fetal surgery for spina bifida repair - fetal medicine midwife - neurosurgeon - fetal neuroradiologist - psychologist / counsellor <p>Attendance at the meeting should be recorded</p>	Other staff involved in the management of the care may attend in addition to those specified.	Operational Policy	Self declaration	1.3.5	safe, effective,
004	There are agreed pathways in place	There must documented pathways in place for referral, assessment, treatment,	The pathways should specify how the	Operational Policy	Self declaration	1.3.5	safe, effective,

		discharge and follow up, as detailed in the service specification, agreed between the fetal surgery centres and fetal medicine units.	different organisations and groups of professionals should interact at defined stages of the patient journey., and should be reviewed at least every two years Where relevant, pathways should take into account nationally or internationally agreed guidance and standards.				
005	There are agreed clinical guidelines in place as detailed within the service specification.	There are agreed clinical guidelines in place as detailed within the service specification.	Where relevant, guidelines should take into account nationally or internationally agreed guidance and standards.	Operational Policy	Self declaration	1.3.5	safe, effective,

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