SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No:	170106S
Service	Open fetal surgery to treat fetuses with open spina bifida
Commissioner Lead	
Provider Lead	*Subject to process*

1. Scope

1.1 Prescribed Specialised Service

This service specification covers the provision of open fetal (in-utero) surgery and supporting medical services by up to two Fetal Surgery Centres (FSCs) for congenital open spina bifida, the main forms of which are myelomeningocoele and myeloschisis. The service is to be delivered in a shared care pathway together with existing local maternity units / Regional Fetal Medicine Units (RFMUs) and regional neurosurgery centres.

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 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 covers the cost of activity undertaken by FSC 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 provides prenatal surgery for women with a fetus affected by open congenital spina bifida, where both the woman and her fetus meet the inclusion and exclusion criteria to ensure that there is clear evidence of benefit to the fetus compared with the alternative management of post-natal surgery. Local maternity units will refer a women carrying a fetus suspected of having spina bifida to a RFMU for assessment and onward referral to one of the FSCs. The FSC will provide assessment, diagnosis, counselling and open pre-natal surgery by a multi-disciplinary team (MDT) which is experienced in the management of this condition. FSC teams will invite the relevant local maternity unit / RFMU and paediatric neurosurgeons to join discussions and, following diagnosis and the setting out of a treatment plan, shared care with the relevant RFMU and local unit will then take place. The implementation and delivery of this specification by FSCs / local units must include the development of clear written guidance and shared care agreements. Women are to be informed that delivery of all subsequent pregnancies is to be by a planned Caesarean section around 37 weeks to avoid rupture of uterus during labour due to the scar on the uterus from the open fetal surgery procedure.

Each FSC must:

1. have an expert MDT, (see personnel list below) which takes into account the individual circumstances for each woman and her fetus. The clinical lead is to be a fetal medicine specialist (an obstetrician trained to undertake uterine surgery with expertise in open fetal surgery for spina bifida repair;
2. have an appropriate caseload to maintain expertise in the treatment of the condition;
3. provide a high quality expert service which reviews and co-ordinates the delivery of care with referring RFMU, and ensures that referring staff in local maternity units and RFMUs are aware of:
    a. the proposed care plan for diagnosis and treatment by the FSC and the post-surgical support, and
    b. management needs of the patient in the remaining gestational weeks;
4. provide robust patient support services.

The FSCs will operate a joint, final decision-making MDT to ensure equitable access and consistency of clinical decision making and surgical approach. It is expected that the FSC will directly relate to an existing centre of experience and expertise.

Components of the FSC MDT:

1. **Assessment MDT to agree on suitability for surgery:** Core team - fetal medicine specialist who is an obstetrician with training in uterine surgery and in undertaking open fetal surgery for spina bifida repair; fetal medicine midwife; paediatric neurosurgeon; counsellor; fetal neuroradiologist or paediatric radiologist with knowledge and skills of the developing brain and the early diagnosis of brain anomalies to review MRI.
2. **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.
3. **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.
4. **Recovery staff:** Post-operative recovery staff.

Follow up requirements following in utero surgery until the baby is born: The woman will attend the RFMU for regular post-surgical follow up assessment as stipulated in **Appendix 2**: Pathways to ensure the ongoing safety of the pregnancy and, due to the risk of preterm birth, may be advised to stop work after fetal surgery and restrict daily activities until the baby is born.

Inclusion criteria - Mother:

- Singleton fetal pregnancy;
- No serious maternal medical complications;
- Maternal cervix long and closed;
- Known maternal Human Immunodeficiency Virus (HIV), Hepatitis B virus, Hepatitis C virus 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 contraindicates surgery e.g. trisomy 18);
- Fetal kyphoscoliosis less than 30 degrees;
- Fetal MRI confirms 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 less than 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 increases 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.

Patient Care pathway: See **Appendix 2** for flowcharts for:

1. Pre-operative pathway;
2. Peri-operative Pathway;
3. Post-operative Pathway (Inpatient);
4. Post-operative Pathway (Outpatient);
5. Post-natal Pathway (Neonatal);

and see **Table 1** below for description of the care pathway.

Table 1: Description of Patient Care Pathway Steps	
Steps	**Sub-steps**
A) Initial diagnosis (local hospital)	1. Suspected isolated congenital open spina bifida at fetal anomaly ultrasound scan. 2. Provision of patient information, with the same level of information provided about the three options (post-natal surgery, termination and pre-natal surgery) to ensure that all women who have a diagnosis have the same level of information and support to enable them to make an informed decision as to whether to continue with the pregnancy. The fetal medicine team will avoid raising expectations regarding the potential for pre-natal surgery in women who are not eligible.
B) Local maternity unit referral to Regional Fetal Medicine Unit (RFMU) after identification of congenital open spina bifida	1. Review within five days for initial counselling by local fetal medicine specialist. 2. Ultrasound scan 3. Diagnosis 4. 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	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 must 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	1. RFMU undertakes education of the woman (and family) about neural tube defects, community resources, provides initial information regarding prenatal and post-natal 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 post-natal period.
F) RFMU counselling on options for fetus	1. RFMU arranges counselling by regional paediatric neurosurgeon if available, neonatologist; fetal medicine midwife; and maternal-fetal medicine specialist. 2. Counselling to include: • discussion of termination of pregnancy (ToP) as part of informed choice; • surgery – including that there are pre and post-natal surgery options depending on the clinical presentation of the fetus provide patient information leaflet about fetal surgery; • Risk of aneuploidy – offer invasive prenatal diagnosis to provide a genetic diagnosis to rule out syndromic congenital abnormality which contraindicates surgery (e.g. trisomy 18); 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, amniocentesis)	1. If diagnosis confirmed and woman consents, then MRI and amniocentesis plus karyotype or microarray. 2. Review of results of these investigations, including reporting of MRI by paediatric / fetal neuroradiologist. 3. If above criteria met, complete referral to FSC if the woman is interested.
H) RFMU referral to Fetal Surgery Centre (FSC)	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 (plus local unit if appropriate) and receiving FSC to discuss the case, (or outpatient consultation if appropriate). 4. If receiving FSC accepts, proceed to complete referral paperwork.
I) FSC assessment to confirm fetal and maternal condition suitable for open fetal surgery AND FSC detailed current counselling specific to prenatal surgery for congenital open spina bifida	On the same day, the following happens: 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 previous MRI at RFMU not sufficient or if not already performed locally or regionally. Purpose of MRI is to document hindbrain herniation and evaluate any central nervous system anomalies not associated with spina bifida. 4. Review of fetal MRI by neuroradiologist. 5. Review of maternal history, fetal karyotypic findings. 6. Discussion of the woman's condition and the fetal presentation.by the joint FSC Unit final surgical decision-making MDT. 7. 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; neonatologist, paediatric neurosurgeon, obstetric anaesthetist with expertise in fetal anaesthesia (including undertaking pre-operative assessment and fetal medicine midwife. 8. Explain risks including 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. 9. Explain the current outcomes evidence for prenatal surgery and local data. 10. Explain steps during fetal surgery for the fetus and the woman and aftercare for both including restrictions on the woman's activity and likelihood of the need to take early maternity leave. 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, post-natal surgery or ToP.
J) FSC – pre-surgical follow up by telephone or in person depending on logistical considerations	1. Woman has sufficient time to discuss with family and local RFMU and / or local obstetric team about fetal surgery option, and consider for at least 24 hours. 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	1. The day before surgery, a repeat ultrasound is performed to get the accurate estimated fetal weight, and for 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. Under general anaesthetic and epidural anaesthesia, a maternal laparotomy and a hysterotomy is performed under ultrasound guidance to determine the optimum incision. The fetus receives anaesthesia by intramuscular injection. The spinal defect is positioned in the hysterotomy and closed surgically by a paediatric neurosurgeon with experience of post-natal and prenatal spina bifida closure. The fetal heart is monitored by ultrasound with recourse to fetal resuscitation if necessary. The uterus and abdomen is then closed. 4. The woman remains in the FSC high dependency unit for one to two days then returns to the FSC antenatal ward, as clinically required. 5. Ultrasound surveillance of uterine scar for bleeding and amniotic fluid leakage. 6. Post-surgery, the FSC contacts the referring RFMU and arranges the date of transfer of care of the management of the woman back to the RFMU. 7. FSC provides management advice to the regional NHS referring centre neurosurgery department regarding post-delivery care, (copied to RFMU and local maternity unit for information). 8. Woman goes home.

L) RFMU follow up during remainder of pregnancy	1. RFMU manages care including follow up ultrasound scans and MRI according to FSC protocol to include documentation of reversal of 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 communicates with receiving FSC to seek advice.
M) RFMU Delivery and NICU care	1. Delivery is by Caesarean section by an experienced obstetric surgeon who will inspect the hysterotomy scar, repairing the defect if needed. 2. Immediate post-natal care for the baby takes place in the regional Neonatal Intensive Care Unit (NICU). 3. If the membranes rupture or the woman labours, the woman presents 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. 4. An individualised care plan is put in place which takes into account risk of preterm labour, infection, prematurity and other maternal and fetal factors. 5. If labouring, woman goes to the nearest maternity centre, which refers her up to the RFMU or calls the RFMU for telephone advice if time does not permit for transfer. 6. Note in clinical notes that all subsequent maternal deliveries must 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	1. Regional NHS referring neurosurgery centre and neonatal departments jointly organise post-natal follow up of the infant, including enhanced developmental support and surveillance. 2. Follow up is at least up to two years of age, corrected for prematurity as per NICE guidelines for babies born preterm (less than 37 weeks) if they are considered at high risk of problems and up to age four for babies born at less than 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 paediatric neurosurgeons.

2.2 Interdependence with Other Services

Clinical co-dependencies include:

- **Local Maternity / Paediatric Unit:** Obstetrics and Paediatrics / Neonatology. Onward referral to neurology and neurodisability as required.
- **Regional neurosurgery units:** Although outside the scope of this service specification, regional neurosurgery units are expected to have access to ongoing pathways of care which include neurological and neurodisability services, urology (bladder management), orthopaedic, physiotherapy and psychological support as required with provision for transition to adult and lifelong care.
- **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 has the following in place:**
  - **a) Infrastructure:** Co-located (all facilities in one unit): fetal surgery clinic capacity; fetal MRI neuroradiology; Level 3 neonatal intensive care unit (NICU); obstetric high dependency unit; maternity unit and inpatient ward for the care of pregnant women which is staffed by midwives to monitor fetus and nurses; fetal medicine theatre lists; theatres; Level 3 adult intensive care unit (which should only be required in exceptional circumstances).
  - **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; Paediatric / Fetal MRI neuroradiology expertise; adult critical care.

3. Population Covered and Population Needs

3.1 The population covered is all women carrying a fetus suspected of having spina bifida who fall within the direct commissioning responsibilities of NHS England who meet the inclusion and exclusion criteria.

This service specification covers the population defined as the commissioning responsibility of NHS England. Commissioning arrangements for the devolved nations in relation to this service are as set out in *UK-wide Commissioning Arrangements of Highly Specialised Services*.

Trusts performing procedures on patients outside of S2 arrangements and aligned referral arrangements need to continue to make financial arrangements directly with the governments involved, separately from their contract with NHS England.

3.2 Population Needs

Approximately 700 women per year in the UK have an ultrasound scan performed in the first half of pregnancy that diagnoses their fetus as having a major form of open spina bifida (myelomeningocoele or myeloschisis), where the spinal column and cord are not properly formed. 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. 100 babies are born in England each year with spina bifida, not all of whom are suitable for prenatal surgery. Data from current European Open Fetal Surgery Centres suggest that only 20% of women with an affected fetus choose to undergo fetal surgery treatment meaning that they might operate on around 10 to 20 cases per year from the UK, including up to 10 from England.

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 women and their partners is particularly important.

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 controlled trial (RCT) in the USA (MOMS) of open surgery 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. The morbidity of open spinal lesions is dependent primarily on the proximal level of the lesion and the Chiari 11 malformation (Ovaere 2014). See **Appendix 1** for Map of the Spine / Vertebral Column.

- 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
- 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 post-natal 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 post-natal 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 post-natal group (P less than 0.0001). Actual rates of shunt placement were 44% and 84% in the two groups respectively.
- Werner et al (2012) Prenatal myelomeningocoele repair is cost effective and frequently cost saving compared with post-natal myelomeningocoele 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 less than 0.001) [as compared to post-natal 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 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 are to 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. See Schedule 4D for CQUIN goals.

Table 2: NHS Outcomes Domains Indicators				
	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 24 months to 36 months gestationally corrected age.	Provider	1,3,5	safe, effective
116	Percentage of babies with improvement in Chiari II malformation.	Provider	1,3,5	safe, effective
117	Percentage of babies having surgery for infantile Chiari II malformation within 12 months of delivery.	Provider	1,3,5	safe, effective
118	Percentage of women offered an appointment by the FSC within one week of referral by the RFMU.	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 FSC MDT.	Self declaration	4	caring, responsive

Structure and Process				
001	There is a specialist FSC 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 FSC multidisciplinary discussion of each referral to agree the diagnosis and treatment plan.	Self declaration	1.3.5	safe, effective
004	There is a joint, surgical MDT across the commissioned FSCs.	Self declaration	1,3,5	Safe, effective
005	There are agreed pathways in place.	Self declaration	1.3.5	safe, effective
006	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

Providers must 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; 'Developmental follow-up of children and young people born preterm' which can be found at: https://www.nice.org.uk/guidance/NG72.

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. It is a requirement to hold national annual clinical meetings involving all commissioned FSCs on an annual basis to include NHS England highly specialised commissioners. FSCs must contribute to national data collection of treatment and outcomes and support development of a national database. Each FSC must ensure that: all practitioners participate in continuous professional development and networking; patient outcome data are recorded and audited across the service; it participates in the national annual clinical outcomes process commissioned by NHS England, including the collection of experience and outcome data from their Hub and Referrers.

Annual Clinical Meetings must address: Clinical performance and outcomes; Process-related indicators e.g. efficiency of the assessment process, prescribing policy, bed provision and occupancy, outpatient follow-up etcetera; Stakeholder satisfaction, including feedback from patients, their families, referring clinician and GPs. Although the provision of post-natal care is outside of the remit of this specification, there is an expectation that local / regional services provide an MDT approach on long term follow-up and that local / regional units provide the FSC with data to support long term outcomes for this service.

5.3 Other Applicable Local Standards

Not applicable.

6. Designated Providers (if applicable)

Not applicable.

7. Abbreviations and Terminology Explained

Chiari II malformation	A condition where the lower part of the brain pushes down into the spinal canal https://www.nhs.uk/conditions/chiari-malformation/ (see also spina bifida aperta). Also known as hindbrain herniation.
CSF	Cerebrospinal fluid
Fetus	The term used for the baby whilst it is still in its mother's womb / uterus. It is only called a baby after birth.
Fetal surgery	Surgery performed on a fetus whilst it is still in the womb / uterus.
FSC	National Fetal Surgery Centre

Hindbrain herniation	This is where the lower part of the brain pushes down into the spinal canal.
Hydro-cephalus	Also known as ventriculomegaly - the brain is surrounded by a clear fluid called cerebral spinal fluid (CSF). In spina bifida, 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.

Karyotype or microarray	Forms of genetic testing
LMU	Local Maternity Unit
MCUG / MCU	Micturating cystourethrogram
MDT	Multi-disciplinary team
MRI	Magnetic Resonance Imaging
MOMS	Management of Myelomeningocoele clinical trial https://www.ncbi.nlm.nih.gov/pubmed/21306277
NICU	Neonatal Intensive Care Unit - a unit commissioned to provide intensive care for newborn babies as set out in the Neonatal Critical Care service specification E08/S/a see https://www.england.nhs.uk/commissioning/wp-content/uploads/sites/12/2015/01/e08-serv-spec-neonatal-critical.pdf
Open spina bifida	The spinal cord and nerve tissue bulge through an open hole or area of thin membrane on the baby's back. This form of spina bifida is also called meningomyelocele or myelomeningocoele.
Open fetal surgery	Whilst the baby is still in the womb, the woman will undergo a surgical procedure where her skin and then her uterus are opened using a surgical incision - this will be slightly wider than the cut used for a caesarean section but is in the same place in the abdomen. The uterus (womb) is then opened to allow the surgeons to access the baby. The spina bifida defect is examined and surgically closed by the paediatric neurosurgeon. The woman's uterus and abdomen are then both closed. The baby will then remain in the womb until approximately 37

	weeks, when it will be delivered in the woman's regional fetal medicine unit by caesarean section.
PTL	Preterm Labour (premature).
RFMU	Regional Fetal Medicine Unit – this term is used in the specification to refer to the designated centre or unit which usually has the most expert role in a geographic area in assessing, diagnosing and counselling women with a fetus suspected of having spina bifida. It is recognised that occasionally, a LMU has long-standing expertise in the diagnosis, investigation, counselling and post-natal of spina bifida. In those circumstances, the LMU advises the RFMU of the woman's circumstances, there is agreement on referral to the FSC and there is ongoing work in a shared care model.
RCT	Randomised Control Trial
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 meningomyelocele or myelomeningocele. These defects are often associated with the Chiari II malformation.
SROM	Spontaneous rupture of the membranes.
ToP	Termination of pregnancy.
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.

References:

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

Cohen. A.R., Couto, J., Cummings, J.J., Johnson.A., Joseph.G., Kaufman.B.A., Litman.R.S., Meanrd.M.K, Moldenhauer.J.S., Pringle.K.C., Schwartz, M.Z., Walker.W.O jr., Warf.B.C., Wax. J.R., MMC Maternal Fetal management Task

Force. (2014) Position Statement on Fetal Myelomeningocele Repair, Am J Obstet Gynael, Feb; 210(2): 107-11.

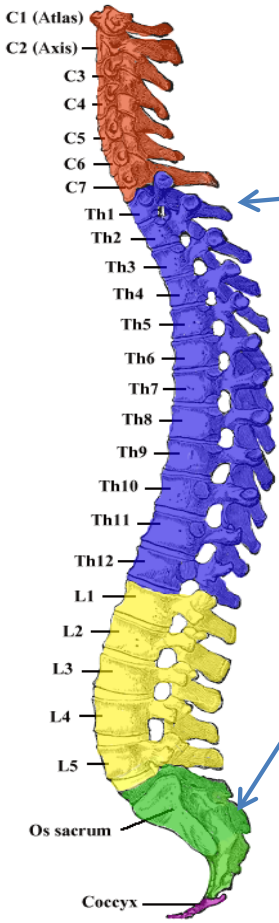
Ovaere, C., Eggink, A., Richter, J., Cohen-Overbeek, T.E, Van Calenbergh, F., Jansen.K., Oepkes, D., Devlieger, R., De Catte, L., Deprest, J.A.; (2014): Prenatal Diagnosis and Patient Preferences in Patients with Neural Tube Defects around the Advent of Fetal Surgery in Belgium and Holland Patients with Neural Tube Defects around the Advent of Fetal Surgery in Belgium and Holland. Fetal Diagnosis and Therapy. DOI:10.1159/000365214.

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

Werner, E.F, Han, C.S, Burd, I, Lipkind, H.S., Copel, J.A, Bahtiyar, M.O., Thung, S.F. (Ultraound Obstet Gynaecol 2012; 40: 158-164), Evaluating the cost-effectiveness of prenatal surgery for myelomeningocele: a decision analysis.

Appendices

Appendix 1: Map of Spine / 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.

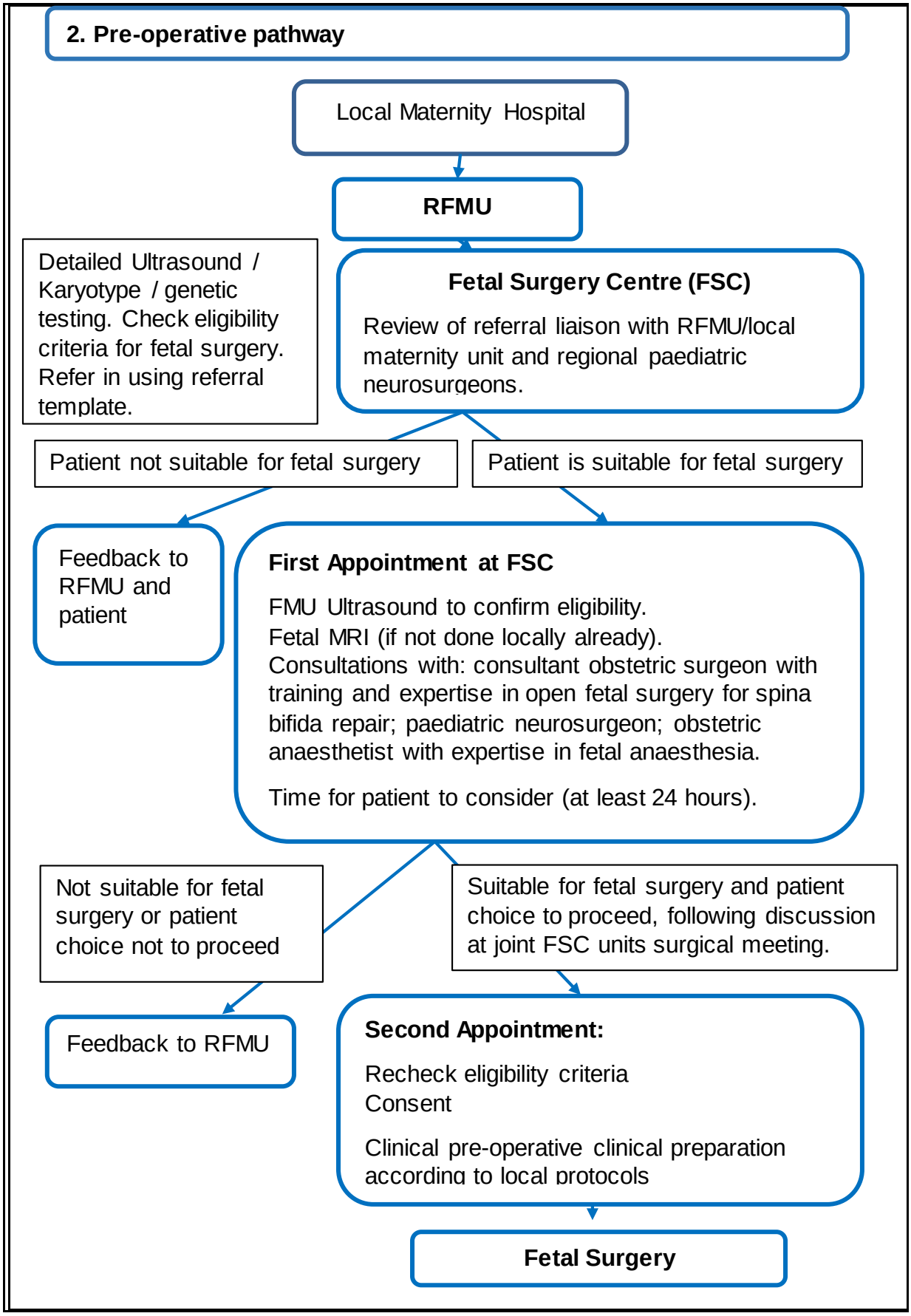
Only spinal lesions in fetuses that occur anywhere between levels T1 to S1 are in scope for open pre-natal spina bifida surgery.

Appendix 2: Patient Care Pathways showing steps

1. Pre-pathway discussions with woman at local maternity unit

- Review within five days for initial counselling by local fetal medicine specialist.
- Ultrasound scan
- Diagnosis
- Provision of patient information with the pregnancy.

Make onward referral to RFMU for confirmation of diagnosis and to discuss options.


2. Pre-operative pathway
Local Maternity Hospital
RFMU
Detailed Ultrasound / Karyotype / genetic testing. Check eligibility criteria for fetal surgery. Refer in using referral template.
Fetal Surgery Centre (FSC)
Review of referral liaison with RFMU/local maternity unit and regional paediatric neurosurgeons.
Patient not suitable for fetal surgery
Patient is suitable for fetal surgery
Feedback to RFMU and patient
First Appointment at FSC
FMU Ultrasound to confirm eligibility.
Fetal MRI (if not done locally already).
Consultations with: consultant obstetric surgeon with training and expertise in open fetal surgery for spina bifida repair; paediatric neurosurgeon; obstetric anaesthetist with expertise in fetal anaesthesia.
Time for patient to consider (at least 24 hours).
Not suitable for fetal surgery or patient choice not to proceed
Suitable for fetal surgery and patient choice to proceed, following discussion at joint FSC units surgical meeting.
Feedback to RFMU
Second Appointment:
Recheck eligibility criteria
Consent
Clinical pre-operative clinical preparation according to local protocols
Fetal Surgery

3. 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

- Ensure preparations complete 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.

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

See postoperative pathway

4. Post-operative Pathway (Inpatient)

Day 0: Close Observation Bay / High Dependency Unit

- Routine post-operative 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

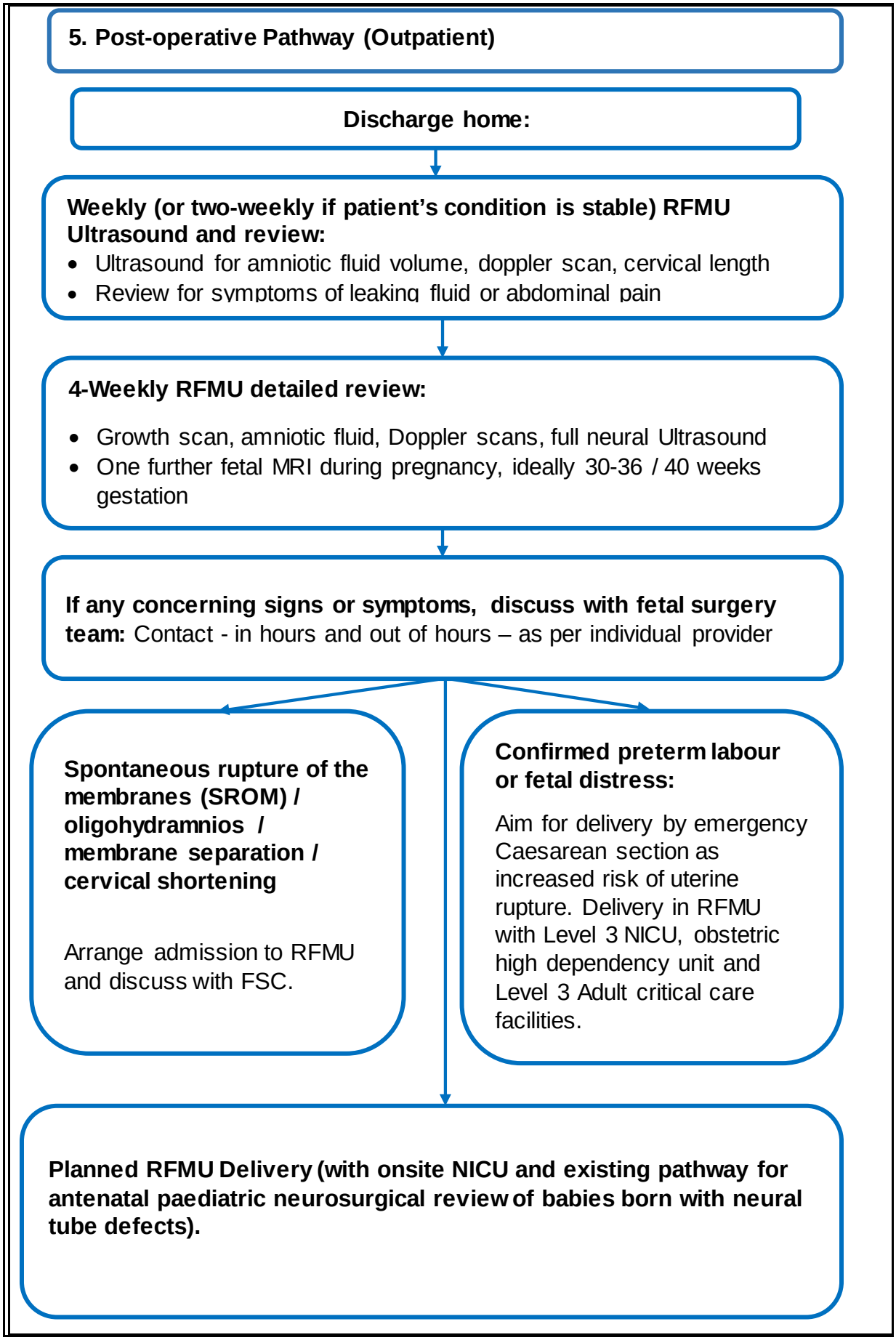
Day 1 - 2: Close Observation Bay / High Dependency Unit

- 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 woman 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. Comprehensive post-surgery discharge letter will be provided to the local unit and RFMU on the care of the woman and her fetus / baby as to any particular management steps to be followed.


5. 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 as increased risk of uterine rupture. Delivery in RFMU with Level 3 NICU, obstetric high dependency unit and Level 3 Adult critical care facilities.
Planned RFMU Delivery (with onsite NICU and existing pathway for antenatal paediatric neurosurgical review of babies born with neural tube defects).

6. Postnatal Pathway (Neonatal)

Immediate:

Admission to regional NICU

Day 0:

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

Appendix 3 Full 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			Provider	1,3,5	safe, effective

		Number of women having fetal surgery.					
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			Provider	1,3,5	safe, effective

		number of fetuses having surgery.					
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 number of fetuses having surgery.			Provider	1,3,5	safe, effective
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 30 months.	Numerator number of babies with recorded assessment of functional motor and sensory level at 30 months Denominator number of births following fetal surgery.			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 24 months to 36 months gestationally corrected age.	Numerator number of babies with recorded complete results of a structured assessment for neurodisability (cognitive and impaired movement) between 24 months to 36 months gestationally corrected age. Denominator number of births following fetal surgery.			Provider	1,3,5	safe, effective
116	Percentage of babies with improvement in Chiari II malformation.	Numerator number of babies with improvement in Chiari II malformation			Provider	1,3,5	safe, effective

		Denominator number of births following fetal surgery.					
117	Percentage of babies having surgery for infantile Chiari II malformation within 12 months of delivery.	Numerator number of babies having surgery for 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 within one week of referral by the Regional Fetal Medicine Unit.	Numerator number of women offered an appointment by the Fetal Surgery Centre within one week of referral by the Regional Fetal Medicine Unit Denominator number of women referred.			Provider	1,3,5	safe, effective

	Patient Experience						
201	There is information for women and families.	The service must have written information for women and families covering at least the following: - information about the service including names and functions / roles of the specialist team - description of the pathway The information has been approved - 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.	The information must be agreed through the Trust's 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.	Operational Policy.	Self declaration	4	caring, responsive
202	Feedback from women and families is reviewed and informs service development and improvements.	The service must have undertaken an exercise to obtain feedback on the women's and families' experience of the services		Annual report	Self declaration	4	caring, responsive

		offered. The exercise and results are presented to and discussed at divisional or directorate level.					
203	Women and families receive counselling by the FSC MDT.	Women and families must be given counselling by members of the FSC MDT including obstetric surgeon with training in fetal surgery and expertise in open fetal surgery for spina bifida repair, neonatologist, paediatric neurosurgeon, obstetric anaesthetist and fetal medicine midwife prior to surgery.		Operational policy	Self declaration	4	caring, responsive
	Structure and Process - infrastructure requirements, staffing, facilities etc						
001	There is a specialist FSC multidisciplinary team.	There is a specialist FSC multidisciplinary team which includes: - 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		Operational Policy	Self declaration	1.3.5	safe, effective,

		- 2 obstetric anaesthetists with expertise in fetal anaesthesia - obstetrician - psychologist / counsellor all above have experience in fetal surgery and have this specified within their job plans / timetable					
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 for fetal open spina bifida.		Training programme	Self declaration	1.3.5	safe, effective,
003	The FSC has a multidisciplinary discussion for each referral to agree the diagnosis and treatment plan.	The FSC has a multidisciplinary discussion for agreeing the diagnosis and treatment plan of women referred for fetal surgery. The discussion includes at least the following: - fetal medicine specialist - obstetric surgeon with training in fetal surgery and expertise in open fetal surgery for spina bifida repair - fetal medicine midwife - paediatric neurosurgeon - fetal neuroradiologist - psychologist / counsellor	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,

		Attendance at the meeting is recorded					
004	There is a joint, surgical MDT across the commissioned FSCs.	All patients referred for surgery are discussed at a joint surgical MDT meeting that includes representation from commissioned FSCs. The joint MDT should comprise at least: paediatric neurosurgeons from the commissioned FSCs; fetal medicine specialists from the commissioned FSCs; other staffing groups may also be invited as deemed appropriate. Treatment decisions and decisions at meetings are recorded.	TBA	Operational Policy	Self declaration	1.3.5	safe, effective,
005	There are agreed pathways in place.	There must documented pathways in place for referral, assessment, treatment,	The pathways must specify how the different	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.	organisations and groups of professionals must interact at defined stages of the patient journey and must be reviewed at least every two years. Where relevant, pathways must take into account nationally or internationally agreed guidance and standards.				
006	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 must take into account nationally or internationally agreed guidance and standards.	Operational Policy	Self declaration	1.3.5	safe, effective,

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