




SFM Fetal Therapy Practice Guidelines: Open Fetal Surgery

Reema Bhatt¹  Vivek Krishnan²  Anusmita Saha³ 

¹Department of Fetal Medicine and Perinatology, Amrita Institute of Medical Sciences and Research Centre, Faridabad, Haryana, India

²Department of Fetal Medicine and Perinatology, Amrita Institute of Medical Sciences and Research Centre, Kochi, Kerala, India

³Department of Fetal Medicine, Amrita Institute of Medical Sciences and Research Centre, Faridabad, Haryana, India

Address for correspondence Reema Bhatt, MS, DNB, FICOG, MNAMS, Department of Fetal Medicine and Perinatology, Amrita Institute of Medical Sciences and Research Centre, Faridabad 121002, Haryana, India (e-mail: id-reemakumarbhatt@gmail.com).

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Abstract

Keywords

- ▶ congenital diaphragmatic hernia
- ▶ fetal lung masses
- ▶ fetal spina bifida utero repair
- ▶ lower urinary tract obstruction/bladder outlet obstruction
- ▶ open fetal surgery

Open fetal surgery allows for surgical intervention on the fetus while maintaining placental perfusion, with the objective of prolonging intrauterine gestation as long as feasible after an operation. Fetal spina bifida utero repair, congenital diaphragmatic hernia, lower urinary tract obstruction/bladder outlet obstruction, and fetal lung masses are the pathologies that can be treated with open fetal surgery. With the availability of minimally invasive techniques, the open technique is not the primary choice of treatment; however, it is prominently used for meningocele repair.

Indications

1. Fetal spina bifida repair
2. Congenital diaphragmatic hernia (CDH)
3. Fetal lung masses
4. Bladder outlet obstruction
5. Congenital Pulmonary Adenomatoid Malformation (CPAM) (lobectomy)
6. Sacrococcygeal teratoma

Consent

Detailed consent should be obtained from the parents, explaining the risks involved to the mother and fetus. The risks for the mother include:

1. Risk of general anesthesia
2. Intrapartum and postpartum hemorrhage

3. Extension of the incision line
4. Need for massive blood transfusion
5. Need for intensive care unit care
6. Peripartum or postpartum hysterectomy
7. Pulmonary edema
8. Sepsis
9. Disseminated Intravascular Coagulation (DIC)
10. Shock

The Risks for the Fetus Includes

1. Hypotension
2. Hypoxia
3. Perinatal death if the airway cannot be secured
4. Fetal brain damage
5. Need for further definitive surgeries

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Perioperative Considerations for Open Fetal Surgery

Prenatal Evaluation

1. Referral to a tertiary care center with an expert team
2. Accurate prenatal diagnosis/precise diagnostic imaging and evaluation for proper planning and management
3. Complete prenatal workup as necessary: e.g., genetic testing, amniocentesis, fetal echocardiography, and magnetic resonance imaging
4. Formation of an experienced multidisciplinary team and operative planning
5. In-depth multidisciplinary prenatal education and counseling for the couple

Preoperative Evaluation By

1. Maternal–fetal medicine expert
2. Pediatric surgeon
3. Pediatric neurosurgeon
4. Anesthesiologist
5. Neonatologist

Intraoperative Team

1. Maternal anesthesiologist
2. Fetal anesthesiologist
3. Scrub nurses-2
4. Circulating nurses-2
5. Airway cart nurse
6. Maternal–fetal medicine specialists
7. Pediatric surgeons
8. Obstetrician
9. Extracorporeal membrane oxygenation

Neonatal team: (In case of preterm delivery)

1. Neonatologist
2. Surgical advanced practice nurses

Essential elements of open procedure.

• Uterine relaxation

1. Inhalational agents
2. Tocolytic agents

• Maintenance of uterine volume

1. Amnioinfusion
2. Partial delivery

• Fetal

1. Anesthetic
2. Blood product availability
3. Echocardiography

4. Intravenous access
5. Pulse oximetry

• Maternal

1. Arterial line
2. Blood product availability
3. Electrocardiogram
4. End-tidal carbon dioxide
5. Pulse oximetry

Maternal Operation Theater (OT) Setup

Maternal Instrument Table

1. Cesarean delivery instrument set and drapes
2. Cord clamp
3. Surgeon-specific sutures, gloves, and supplies

Preoperative Checklist

1. Surgical consent
2. Review patient's medical history, physical examination, imaging
3. Presence of allergies
4. Review of the patient's baseline vital signs
5. Confirm the patient's identity
6. The planned surgical procedure
7. Preoperative medications

Operating Room Requirements

1. Placement on the OT table includes supine positioning with a leftward tilt to prevent compression of the inferior vena cava.
2. Adequate intravenous access is established and sequential compression devices are placed.
3. A preoperative ultrasound is performed to define the placenta and assess the position of the fetus. This helps with the decision of hysterotomy location to avoid bleeding or other complications. The planned site of the incision is planned.
4. Maternal, deep general anesthesia is then established, ensuring adequate doses of inhalation anesthetics to maintain uterine relaxation throughout the procedure, avoiding maternal hypotension. Indirect fetal anesthesia is achieved.
5. The fetus is also administered intramuscular anesthesia in the thigh that includes fentanyl, pancuronium/atracurium, and atropine combination.
6. Adjunctive myometrial tocolytics that are used include atosiban or terbutaline. Magnesium sulfate is also used in some centers.
7. Invasive maternal blood pressure monitoring is performed in certain centers due to the risk of hypotension in the setting of high-dose anesthetics, impaired venous return, and the potential for large-volume hemorrhage.
8. It is essential to maintain adequate maternal blood pressure to provide appropriate uteroplacental circulation and subsequent fetal perfusion during the procedure.

9. Other considerations prior to proceeding with the operation include placement of a urinary catheter and epidural catheter for postoperative maternal pain management

Procedure Steps

1. A laparotomy incision is made, generally via a Pfannenstiel, Maylard, or low midline incision depending on the anticipated hysterotomy location.
2. Ultrasound is used to map placental borders and select the optimal hysterotomy site. A hysterotomy is made away from the placenta generally in the lower uterine segment to reduce blood loss. In the case of placenta previa or anterior placenta, the incision may be in the upper segment or fundal. Special hysterotomy staplers may be used to avoid blood loss. The placement of uterine sutures at the margins of planned uterine incisions to decrease blood loss is considered by some surgeons.
3. Fetal hemodynamic monitoring is established and medications are administered to provide analgesia and paralysis via either the intramuscular or intravenous route (usually transplacental anesthesia works).
4. At the same time, the team can proceed with the fetal intervention as planned.
5. The fetus is positioned according to the planned intervention which in meningocele repair requires the spine to be anterior, in which case some manipulation is needed to adjust the fetal position.
6. In meningocele, repair involves the surgical covering of lesions above S1 between 19 and 25 completed weeks of gestation. This leads to better results than postnatal repair or in utero repair of the lesion where the prenatal defect closure approach is equivalent to the postnatal surgical process.
7. The “MOMS” trial is a randomized controlled trial that compared fetal operative repair with standard postnatal repair. Meningocele (MMC) is repaired by open fetal surgery with the closure of the native dura, myofascial flaps, and skin over the defect. The trial showed significantly superior outcomes in the fetal intervention group. The study found that infants in the surgical group were 50% less likely to require ventriculoperitoneal shunt and had a significant decrease in the incidence of the Chiari malformation. In addition, 42% of infants were walking independently at 30 months compared with 21% in the standard therapy group, a significant improvement. In long-term follow-up, neurological deficits were less severe in those who underwent fetal intervention.
8. Fetal lobectomy or sacrococcygeal teratoma excision can also be done by open fetal surgery.
9. After the surgery is completed, the uterus is closed and warmed Ringer lactate containing antibiotics is injected into the uterus to replace lost amniotic fluid. In some centers, the amniotic fluid is aspirated as the uterine incision is taken that if not contaminated is reinstilled in the uterus after completion of the surgery.

Postoperative Considerations

1. Both the mother and the fetus require intensive monitoring and stringent tocolytic treatment following surgery. Magnesium sulfate is considered a good choice by many centers.
2. Postoperative complications include amniotic fluid leaks, chorioamniotic separation, and noncardiogenic pulmonary edema in the mother.
3. At 36 weeks of gestation, the fetus is normally delivered via scheduled cesarean surgery. If pregnancy-related difficulties arise, an earlier delivery may be necessary.
4. Preterm labor is the most prevalent consequence of open fetal surgery, with a mean gestational age of 34 weeks.

Patient Information Leaflet

• What is the open fetal surgical procedure?

An open procedure is the surgical delivery of your baby through incisions in your abdominal wall and uterus. The repair of the structural defect will be done by a team of experts. Once surgery is performed, the uterus will be closed and pregnancy will be continued till at least 36 weeks. If a patient goes into preterm labor, an earlier delivery may be warranted.

• What are the risk factors for undergoing the procedure?

Only you and your doctor can decide whether the benefits outweigh the risks. Some possible risks for both you and your baby include:

For you

1. Risk of general anesthesia
2. Risk of significant blood loss post partum haemorrhage (PPH), requirement for blood transfusion
3. Risk of incision extension
4. Risk of postoperative infection
5. Risk of uterine rupture in current and future pregnancies
6. Preterm delivery

Fetal risks

1. Hypotension
2. Hypoxia
3. Hypothermia
4. Bradycardia
5. Nerve palsies
6. Fetal/neonatal demise

• Where will the surgery be done?

The surgery may be performed in a hospital that specializes in the care of pregnant mothers. A team of healthcare providers will be there to do your surgery and a separate team will work with your baby.

• What is the postoperative care of the mother after the procedure?

- The postoperative course may be worsened by
- Preterm delivery
- Amniotic fluid leaks

- Chorioamniotic separation
- Development of noncardiogenic pulmonary edema in the mother
- Sepsis
- Uterine rupture
- **What happens after discharge?**
- **Frequent antenatal care (ANC) visits**

Call Your Doctor Immediately If You Experience Any of the Following Postoperative Complications

1. Painful contractions,
2. Excessive vaginal bleeding,
3. Swelling or excessive bleeding from the surgical site,
4. Painful and/or swollen and red calves, or
5. Foul-smelling vaginal discharge.

Other Postoperative Complications Depend on the Procedure Performed

Conflict of Interest

None declared.

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Suggested Reading

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