ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 5, Issue – 1, January - 2022, Page No. : 382 - 388

Let Us Mend the Bend Before Birth- An overview of prenatal repair in Spina bifida

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Citation of this Article: Dr. Chrishantha Joybell, "Let Us Mend the Bend Before Birth- An overview of prenatal repair in Spina bifida", IJDSIR- January - 2022, Vol. – 5, Issue - 1, P. No. 382 – 388.

Type of Publication: Review Article

Conflicts of Interest: Nil

Introduction

Spina bifida is a birth defect that occurs when the spine and the spinal cord does not form properly. It is a type of neural tube defect. The neural tube is the structure in a developing embryo that eventually becomes the baby's brain, spinal cord and the tissues that enclose them. It occurs when a developing baby's spinal cord fails to develop or close properly while in the womb. The worldwide incidence of spina bifida is approximately 4.63 per 10,000 live births.[1] The world-wide variations are recognised as myeloschisis, in which the neural tissue is 'flush' with the surrounding skin; and myelomeningocele (MMC), in which the spinal cord and meninges protrude beyond the plane of the skin as a cerebrospinal fluid (CSF)-filled sac. These two variations are collectively referred to as 'spina bifida' throughout [2].

Signs

The skin above the spinal defect may include an abnormal tuft of hair, a birthmark or protruding spinal cord tissue brown spot-on skin, or skin cyst. It requires a medical diagnosis for detailed evaluation. Clinically, spina bifida leads to difficulties with mobility and ambulation, which is largely dependent on lesion level; by adulthood, independent ambulation is seen in 93% of patients with a sacral lesion, 91% with an L5 lesion, 57% with an L4 lesion and no patients with an L1–3 or thoracic lesion[3].

Symptoms

People may experience, Gastrointestinal symptoms that include bowel obstruction, constipation, or leaking of stool. Muscular: muscle weakness, stiff muscles, or overactive reflexes. Urinary: bed wetting, leaking of urine, or urinary tract infection. Sensory deficits and orthopedic abnormalities, such as talipes(clubfoot), kyphosis and scoliosis can also occur. Most of the patients with spina bifida experience neurogenic sphincter dysfunction resulting in impaired bladder and bowel control, which is commonly managed with a combination of toileting regimens, clean intermittent self-catheterisation and medications such as laxatives, enemas and anticholinergics[4, 5]. Also common are hunched back, intellectual disability, nerve injury, paralysis, paralysis of the body's lower half, or physical deformity.

Spina bifida is also associated with a reduced IQ[6].Although not a lethal condition, spina bifida is associated with a reduced life expectancy and early mortality, particularly in those with higher lesions[7].Having a child with spina bifida is associated with higher levels of parental [8] and sibling[9] stress.

Treatment

When treatment is necessary, it is carried out through surgery to close the defect. Other treatment modalities focus on managing complications.

Spinal surgery

Correcting abnormalities of the spinal cord or its surrounding bones through surgical methods.

Colostomy

Chronic constipation and associated symptoms in adult patients with spina bifida can be treated with a loop colostomy procedure. This procedure is an innovative and helpful option for these specific patients.

Spinal fusion

Mazur et al from his study unveiled the functional status and ambulation after spinal fusion in 49 children with spina bifida. They found deterioration in ambulation in 67% of patients who had undergone combined anterior and posterior fusion, in 57% following anterior fusion, and in 27% following posterior fusion alone.

Suprapubic cystostomy is a procedure to help drain the bladder (organ that collects and holds urine). A tube called a catheter, which leads out of the lower abdomen, is inserted to drain the bladder.

This review article mainly intends to throw light on the importance of prenatal repair, so that when a child is born on this earth, let he or she know that the earth is a beautiful place to live. Though their life is hard and challenging, we do not have the power to make their life fair, but we do have the power to make their life sustainable as much as we can.

Prenatal Repair

Prenatal repair of myelomeningocele (MMC), the most common and severe form of spina bifida, is a delicate surgical procedure where fetal surgeons open the uterus and close the opening in the baby's back while they are still in the womb. Because spinal cord damage is progressive during gestation, prenatal repair of myelomeningocele may prevent further damage.

Open fetal repair of the spinal lesion has been shown to improve hindbrain herniation, ventriculo peritoneal shunting, independent mobility and bladder outcomes for the child and, despite an increased risk of prematurity, does not seem to increase the risk of neuro developmental impairment[2].

Fetal spina bifida surgery is one of the most exciting challenging developments in the history of treatment for birth defects. It is an extremely complex procedure that requires significant commitment on the part of mothers who choose to go forward with it and it requires extensive surgically experienced hands for a high success rate.

Fetal surgery for spina bifida is not a cure, but studies show that prenatal repair can offer significantly better results than traditional postnatal repair. Fetal surgery for spina bifida greatly reduces the need to divert fluid from the brain, improves mobility and improves the chances that a child will be able to walk independently.

Mothers who choose fetal surgery require the most expert and carefully coordinated care from the time of diagnosis, for the spina bifida surgery itself, and through the baby's delivery and care afterwards. Comprehensive counselling is done on the condition and the risks involved in the procedure.

Before proceeding with fetal surgery, you will undergo a physical exam and complete review of your medical history to obtain clearance for surgery. The pre-surgery

consultation will include: A thorough explanation of care and monitoring both before and after delivery, including a review of medications necessary before, during and after fetal surgery. A review of the surgical procedure and its risks, including preterm birth, uterine scarring, membrane separation, infection, bleeding, prematurity and fetal demise. A consultation with a social worker and a psychological evaluation to assess readiness for surgery, coping mechanisms and family support.

Table 1: Inclusion and exclusion criteria for fetal surgery in Management of Myelomeningocele Study (MOMS) trial[10]

Inclusion criteria	Exclusion criteria
Myelomeningocele (including myeloschisis) at levelT1 through S1	Multifetal pregnancy
Maternal age≥18 years	Insulin dependent pre-gestational diabetes (since 2014, this is no longer a contraindication if well controlled
Gestational age of19+0to 25+6weeksof gestation	Kyphosis in the fetus of 30 degrees or more
Normal karyotype	Current or planned cervical cerclage or documented history of cervical weakness
	Placenta praevia or placental abruption
	Short cervix (<20 mm) measured by transvaginal cervical ultrasound
	Obesity as defined by body mass index of 35or greater (since 2014 this has been moved up to 40)
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	Maternal–fetal Rh isoimmunization, Kell sensitization or a history of neonatal alloimmune thrombocytopenia
	Maternal HIV or hepatitis-B status positive
	Known hepatitis-C positivity; if unknown, screening is not required
	Uterine anomaly such as large or multiple fibroids or Mullerian duct abnormality
	Previous surgery in the uterine corpus
	Patient does not have a support person (e.g., husband, partner, mother)
	Inability to comply with the travel and follow-up requirements of the trial
	Patient does not meet other psychosocial criteria (as determined by the psychosocial interviewer using a standardised assessment) to handle the implications of the trial
	Maternal hypertension that would increase the risk of pre-eclampsia or preterm delivery (including,

	but not limited to uncontrolled hypertension, chronic hypertension with endorgan damage and new onset hypertension in current pregnancy)

The multidisciplinary team includes fetal surgeons, neurosurgeons, maternal-fetal medicine specialists, fetal cardiologists, neonatologists, anesthesiologists, sonographers, advanced practice nurses, a psychologist, social workers and coordinators — all focused on the best outcome for the mother and the baby.

Animal models of spina bifida have suggested that open fetal repair is safe and effective[11].The first open fetal spina bifida repair in humans was performed in the year 1998 [12] and over the next 5 years, several observational studies were published with generally positive results regarding the prognosis of the repair.

Preoperative Procedure

Prenatal repair of myelomeningocele is performed between 23 weeks and 25 weeks 6 days gestation. Mothers will be hospitalized with a support person for the surgery and the entire duration of the pregnancy is subjected to close monitoring.

A full, structural fetal ultrasound scan, including neurosonography and echocardiography, is performed to determine lesion level and size, the degree of any spinal kyphosis, the presence of hindbrain herniationand ventricular size and to exclude any additional abnormalities. Magnetic resonanceimaging (MRI) is used as an adjunct to confirm intracranial findings and exclude any undetected abnormalities.

Operative Procedure

An epidural is inserted prior to surgery to allow for postoperative pain management; deep inhalational general anaesthesia is used to reduce uterine contractility and tone. Maternal intravenous antibiotics are given and tocolysis is administered pre-operatively and intra-operatively. In prenatal repair, the mother receives general anesthesia, which also relaxes the uterus and anesthetizes the fetus. Fetal surgeons perform a laparotomy (an incision across the mother's abdomen). A maternal-fetal medicine specialist performs sterile intraoperative ultrasound to map the position of the placenta and the fetus, and the baby's back is rotated into view. The uterus is then opened with a uterine stapling device that pinches off all blood vessels and keeps membranes secured to the muscle.

A pediatric neurosurgeon removes the MMC sac, if one is present, returns the spinal cord to the spinal canal, and closes the surrounding tissue and skin over the defect to protect the spinal cord from exposure to the amniotic fluid. Before the last few sutures of uterine closure, crystalloid fluid is allowed to accumulate and antibiotics are administered into the amniotic cavity. The uterus and the abdominal incision are then closed. During surgery, a fetal cardiologist uses echocardiography to closely monitor your baby's heart. All of these measures are used to ensure the utmost safety of both mother and baby.

post operative procedure

Mothers usually remain in the hospital for three to five days and are on modified bed rest for three to four weeks after surgery to reduce the risk of preterm labor. For the remainder of the pregnancy, follow-up includes weekly visits to the hospital for ultrasound monitoring and routine prenatal care. If labor does not begin sooner, the baby will be delivered by planned cesarean section at 37 weeks. After delivery, the baby will be cared for in the Newborn/Infant Intensive Care Unit (N/IICU).

Mothers who elect fetal repair will also be asked to sign a consent for postnatal follow-up when the child reaches 12 months, 30 months and five years of age. Detailed follow-up not only provides excellent care for the child, it adds to a growing base of knowledge that will benefit future generations of children with spina bifida.

After the baby is born, he will stay in the N/IICU where he will be closely monitored by a multidisciplinary team of specialists who are experienced in caring for babies born with spina bifida. This team includes neurosurgery, urology, orthopaedics, physical therapy and others from the Spina Bifida Program.

Following procedures are carried out during the baby's N/IICU stay:

1. Head ultrasounds on the day of delivery and prior to discharge.
2. MRI to evaluate the baby's brain and spine.
3. Daily head circumference measurement.
4. Renal and bladder ultrasound at 2 days old.
5. Bladder scans with a handheld ultrasound device every four hours for the first 48 hours to estimate the amount of urine in the bladder and whether the baby will require a catheter.
6. Clean intermittent catheterization if the baby's bladder volume is greater than 50 percent of expected volume.
7. Video-urodynamics at 2 days and 2 months of age. A special catheter measures pressure when the bladder is full. A soft catheter in the rectum measures abdominal pressure on the bladder. A uroflo chair measures urine flow rate and time needed to empty bladder.
8. Follow-up includes visits every four to six months until age 2, then annually, with urodynamic testing and renal bladder ultrasounds to ensure kidneys function properly and bladder function is stable.

Follow Up of Child Patient

Table 2

Day 0	1. Wound review and photograph (please also send to fetal surgery team) 2. Neurosurgery review of spinal wound and documentation of neurology 3. Indwelling catheter until MCUG 4. In non-paediatric neurosurgical setting, arrange for review at regional paediatric neurosurgical spina bifida unit
Inpatient	1. Cranial USS and daily OFC 2. Repeat cranial USS prior to discharge and at 2 weeks (earlier if clinical concern) 3. US urinary tract 4. US hips 5. Physiotherapy review and muscle charting 6. VP shunt if required at discretion of team
Discharge from hospital	Prophylactic antibiotics pending MCUG at 6 weeks
Six-week follow up	1. MCUG 2. USS cranial/renal 3. MRI brain and spine before 3 months if feasible
Three-month follow up	1. Natural fill urodynamics 2. DMSA 3. Bayley Scales of Infant and Toddler Development

Neonatal protocol following open fetal surgery for spinabifida. DMSA = dimercaptosuccinic acid radionuclide scan; MCUG =micturating

cystourethrogram; MRI = magnetic resonance imaging; OFC = occipito-frontal circumference; SCBU = special care baby unit; USS = ultrasound scan; VP = ventriculoperitoneal

Future Alternative Procedures

Mini-hysterotomy

A less invasive alternative to the surgical method described is the use of a 'mini-hysterotomy', i.e. a uterine opening with a diameter of less than 4 cm, as opposed to the 6–8cmopening commonly used. Through this opening, a standard multilayer microsurgical repair is performed[13].

Conclusion

Spina bifida is a congenital central nervous system malformation with lifelong physical and mental effects. Open fetal repair of the spinal lesion has been shown to improve short-term outcomes for the child, with the consequent risks of prematurity and maternal morbidity. Further evidence regarding long-term outcomes, fetoscopic repair and alternative techniques is awaited.

As dental surgeons, though it is very challenging, let us create a beautiful world for these special little feet from heaven to imprint their footprints on the sands of time.

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