Management of pregnancy in a woman with spina bifida: A case report*

By Myza C. Espallardo, MD and Judith M. Sison, MD, FPOGS, MD, FPOGS, FPSURPS
Department of Obstetrics and Gynecology, University of Santo Tomas Hospital

ABSTRACT

With advancements in corrective surgery for spina bifida since the 1960s, affected women are now reaching adulthood and achieving pregnancies. However, the implications on reproductive health—especially in pregnancy—are rarely studied.

We are presenting a case of a woman born with spina bifida who has undergone surgical repair and closure of the defect at 4 years old, as well as surgical management for tethered cord at 13 years old. She lived productively into adulthood, became pregnant and delivered vaginally to a healthy baby despite various obstetrical challenges. In the care for this patient, we faced recurrent urinary tract infections, the risk of preterm delivery and the task of coordinated multidisciplinary care to solve dilemmas in decision making.

Through this case report, we were able to share our experience, explore the most recent evidence to support our clinical decisions and hopefully serve as a basis for future clinical practice recommendations.

Keywords: Spina bifida, pregnancy, tethered cord

INTRODUCTION

The average incidence of neural tube defects is 1/1000 births with significant geographic variation. It is high in less-developed countries where risk factors such as maternal diabetes, obesity, lower socioeconomic status, and exposure to certain teratogens during the conception period are relatively common.

A significant milestone has been achieved in preventing neural tube defects during the 20th century. With the advances in corrective surgery for congenital defects of the spinal cord in the past decades, an increasing number of females born with spinal cord lesions has reached childbearing age. These women are presented with special challenges in reproductive health which are rarely described in literature.

Spina bifida is the most common among a group of defects that result from an incomplete closure of the neural tube during embryologic development. The majority of individuals with this malformation are asymptomatic although they may manifest a variable amount of insult to the somatic and autonomic systems, and adversely affect bowel and bladder functions.

There is a scarcity of literature which focused on the effects that spina bifida may have on women's reproductive health. Concerns on sexual function and pregnancy have been raised and their health care providers do not have the benefit of evidence-based research to address these issues. A number of studies have demonstrated that this population has a predilection for medical complications such as urinary tract infections and deep vein thrombosis, and are also at greater risk for preterm labour. With their irregular spinal anatomy, they present obstetrical dilemmas during childbirth, including the mode of delivery and the type of anaesthesia to be provided. For these reasons, constant planning and complex decision making with the healthcare team is warranted.

Little is reported in literature about pregnancy outcomes in this growing population of women. There are only a few studies on this patient population that have been published and these are limited mostly to case reports and case series. And although these studies have contributed to the management of these women during pregnancy, there are no standards of care that exist to guide health care providers in clinical decision making. In light of this lack of data, this case report aimed to describe the multidisciplinary care for a pregnant woman with spina bifida, and the challenges faced during her pregnancy and eventual delivery.

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CASE REPORT

A 26-year-old primigravid woman, at 38 to 39 weeks of gestation, presented in labor and was admitted on March 18, 2018 in a private tertiary hospital for delivery. She has spina bifida and a history of tethered cord syndrome and initially manifesting with bladder dysfunction.

The patient was born to a then 29-year-old G2P2 (2002) mother, via cesarean section, with a lipomeningocele over the sacral area which was surgically removed and repaired when she was 4 years old. Her mother had no history of gestational diabetes or any comorbid condition, but was not able to take folic acid supplementation during conception. During childhood, she had normal cognitive function and was independent in all activities of daily living. Manual muscle testing was 5/5 on bilateral upper and lower extremities, while deep tendon reflexes were normal. She had a sensory deficit over the right lower extremity and neurogenic bowel dysfunction characterized by poor anal sphincter control and constipation, relieved intermittently with use of laxatives.

At 13 years old, the patient had a history of recurrent urinary tract infection (UTI), having 2 bouts of UTI every month despite appropriate antibiotic use. She was admitted on September 2005 for treatment and evaluation of renal function.

Intravenous pyelography revealed urinary retention as evidenced by a marked dilatation of the right pelvocalyceal structures and both ureters up to the area of the ureterovesical junctions, despite having no opaque lithiasis seen in the region and course of the genito-urinary tract. A DMSA scan was also done which showed multiple photopenic defects in both kidneys—more on the right—and a left kidney contributing about 68% of the overall cortical function. In light of the previous history of surgery for a lipomeningocele, the possibility of a bladder pathology was entertained as a cause for the recurrent UTI, resulting to kidney injury.

Magnetic resonance imaging of the lumbosacral spine showed a tethered cord fixed to the posterior wall of the thecal sac at S2 level. This causes hypoxia to the sacral nerve roots and produces a myriad of symptoms including a neurogenic bladder, bowel dysfunction as experienced by the patient. She started intermittent urethral self-catheterization every 6 hours and was prescribed prophylactic urinary antiseptic—Nitrofurantoin 100 mg once daily—and an anticholinergic, Oxybutynin 10 mg once daily, to aid in micturition control.

Video urodynamic studies showed bladder sphincter dyssynergia and hypertonic bladder with a closed bladder neck. This was supported by nerve conduction studies revealing partial, chronic and neurogenic affectation of the S2, S3 and S4 nerve roots. The resulting neuronal dysfunction may explain the voiding dysfunction and the need to do clean intermittent self-catheterization by the patient. Furthermore, this is the tethered cord syndrome which could be due to adhesions incurred from her previous operation.

Video Urodynamic Studies

	11/21/2006 *6 months post bladder reinnervation	4/6/2007 *no more CIC	12/16/2013
Maximum bladder capacity		204 cc	342 cc
End filling pressure		31 cm H2O	40.2 cm H2O
Maximum detrusor pressure		42 cm H2O	42 cm H2O
Bladder compliance		7 cm H2O	8.14 cm H2O
Detrusor sphincter dyssynergia		Yes	Yes
Voided volume	250 cc		216 cc
Residual urine	N/A		126 cc
ACONTRACTILE BLADDER WITH BETTER COMPLIANCE			

Despite appropriate therapeutic and prophylactic antibiotic therapy, there was persistence of recurrent UTI and repeat DMSA scan after 6 months showed a progressively faltering right kidney function which was down to 13%. The patient was then admitted for meningocoelectomy, release of tethered cord and bladder innervation. She was discharged improved after 6 hospital days and resumed clean intermittent self-catheterization.

Prenatal check-ups started at 7 to 8 weeks age of gestation with a total of 8 visits with the attending physician. She had no history of first trimester vaginal bleeding or hypogastric pain, however, a transvaginal ultrasound done at 5 to 6 weeks age of gestation showed subchorionic hemorrhage amounting to 2.03 cc which persisted until 17 to 18 weeks of gestation. She was prescribed dydrogesterone 10 mg tablet and Isoxuprine 10 mg tablet both taken twice daily until subchorionic hemorrhage resolved. Dydrogesterone was continued until the end of second trimester.

At 12 weeks age of gestation, the patient had curd-like vaginal discharge assessed as vaginal candidiasis and was managed with clotrimazole vaginal suppository for 3 days. At this time, there was persistence of the subchorionic hemorrhage for which Isoxuprine 10 mg and Dydrogesterone 10 mg were continued.

This pregnancy was complicated with persistent asymptomatic urinary tract infections. A urine culture was requested during the first consultation and showed growth of Escherichia coli which was sensitive to most oral antibiotic classes. Regular urinalysis was also done on a monthly basis which consistently revealed pyuria and bacteriuria. The patient at this time was also being seen by a urologist who suggested giving antibiotics as prophylaxis or for treatment whenever with symptoms. The patient however did not have episodes of febrile or symptomatic UTI throughout the antenatal period. She adhered to the prescribed antibiotic regimen and clean intermittent self-catheterization every 4 hours, and went on regular follow-up consultations.

Several consultations with anesthesiologists were made by the obstetrician to determine the most appropriate anesthesia plan for the patient. On clinical pelvimetry, there is no noted restriction of pelvic diameters, thus, delivery via the vaginal route was preferred.

The patient presented with regular uterine contractions and passage of bloody show at 38 weeks and 3 days of gestation. Upon admission, the cervix was 2 cm dilated and 70 percent effaced, with an intact bag of water, the fetus in cephalic presentation at station -3. The patient achieved 4 cm cervical dilatation on the third hour of labor and was subsequently admitted to the delivery room for labor analgesia.

Despite the patient's lumbosacral spine deformities, the epidural space was identified and an epidural catheter was successfully placed at L_{3-4} lumbar interspace while the patient was in the left lateral position. The patient experienced satisfactory pain relief throughout her labor, with stable vital signs, and after 3 hours, she attained full cervical dilatation. The obstetrician performed pudendal block for additional labor analgesia. The patient was allowed to push during the second stage of labor and she had an uneventful vaginal delivery to a 3,340-gram baby girl, with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. Autonomic dysreflexia did not occur during labor.

In the puerperium, she resumed clean, intermittent self-catheterization. She made an uncomplicated recovery and was discharged from the hospital.

DISCUSSION

For the past decades, the life expectancy among women with spina bifida has been increasing, along with the clamor for information about what to expect as they transition to womanhood. Being a special population with unique health concerns, there is a need to define and settle issues on reproduction, particularly pregnancy complications and secondary conditions that these women may encounter.

Because of this relative scarcity of literature, we studied this case which mirrored the medical, obstetrical, and social concerns of this growing population. Incorporated in this case report is the evidence base from MEDLINE searches that guided us in giving the optimal obstetric care of this patient.

Management of Antenatal Complications

Since the 1980s, various case series on women with spinal cord lesions, including those specifically focusing on spina bifida in pregnancy, have described problems related to the urinary tract. In particular, infection and problems of accommodation of the growing uterus within the abdomen in the presence of a restrictive spinal deformity have been the common obstetrical challenges. This also includes preterm labor, risk of prematurity and unattended delivery as the main risks for both the mother and the fetus.

Among females with spina bifida who reach adolescent and adulthood, pregnancy with expected urinary complication is common. However, definitions of urinary tract infection (UTI) are heterogeneous and infrequently applied in studies of spina bifida patients. Asymptomatic bacteriuria, urinary incontinence, vesicoureteral reflux and other genitourinary symptoms that can be caused by tethered cord syndrome are commonly diagnosed as UTI and treated with antibiotics. During her childhood, our patient was diagnosed to have vesicoureteral reflux and neurogenic bladder. There was also damage to the upper urinary tract necessitating early intervention and continuous monitoring.

Most European and North American clinical practice guidelines recommend screening for asymptomatic bacteriuria as a routine test during pregnancy. This is because antibiotic treatment of asymptomatic bacteriuria in pregnant women reduce the complication of upper urinary tract infections and preterm labour. However, the benefit and harm of screening vs. no screening has not been clearly established. In a systematic review of publication until February 2016, four randomized controlled trials were identified comparing antibiotics with no treatment or placebo in 454 pregnant women with asymptomatic bacteriuria. The event rates reported by more recent study were not significantly different in terms of preventing pyelonephritis (0 % vs. 2.2 %; OR = 0.37, CI 0.01-9.25, p = 0.515).[1] For our patient, we requested for a urinalysis each trimester, repeated urinalysis after antibiotic treatment, and provided antibiotic prophylaxis against recurrent infection.

Complete Blood Count

	8/12/17	Reference values
Hemoglobin	142 g/L	120 – 160g/L
Hematocrit	0.41	0.37-0.43
RBC Count	4.80 x 10^{12}/L	4.2 – 5.4 x 10^{12}/L
Platelet Count	352 X 10^{9}/L	170-400 x 10^{9}/L
WBC Count	8.3 X 10^{9}/L	4.0 – 10.0x 10^{9}/L
Neutrophils	0.59	0.50 – 0.70
Lymphocytes	0.29	0.25 – 0.35
Monocytes	0.10	0.03-0.06
Eosinophil	0.02	0.02 – 0.04
Basophil	0	0.00 – 0.01

Urinalysis

	8/12/17 (7-8 weeks AOG)	9/11/17 (11-12 weeks AOG)	10/4/17 (14-15 weeks AOG)	1/5/18 (28-29 weeks AOG) *4 days after Cefuroxime	1/26/18 (31-32 weeks AOG)
Color	Yellow	Light yellow	Yellow	Yellow	Light yellow
Transparency	Turbid	Slightly cloudy	Hazy	Slightly turbid	Slightly turbid
pH	8.0	6.5	7.0	Alkaline	7.0
Specific gravity	1.010	1.020	1.025	1.015	1.010
Glucose	Negative	Negative	Negative	Negative	Negative
Protein	Negative	Negative	Negative	Negative	Negative
WBC	175/hpf	44/hpf	16/hpf	6-15/hpf	1-2/hpf
RBC	2/hpf	1/hpf	2/hpf		
Bacteria	5684/hpf	42/hpf	80/hpf		+

In order to promote rational management of urinary tract infections in pregnant patients with spina bifida, there is a need to include more symptoms like fever and diagnostic tests like urine culture aside from routine urinalysis for detecting asymptomatic bacteriuria.[2] Urine culture requested for our patient revealed E. coli sensitive to first line antibiotics like Cefuroxime.

We also gave our patient, a maintenance of dydrogesterone. A systematic review of Carp in 2015 specifically on dydrogesterone showed lower miscarriage rate in dydrogesterone compared with control (10.5% vs. 23.5%) with odds ratio for miscarriage at 0.29 (confidence interval 0.13 to 0.65) or a 13% absolute reduction in the miscarriage rate. The adverse effects were minimal.[3]

Specialized Prenatal Care

To manage the complications of pregnancy in patients with spina bifida, prenatal care needs to be more intensive. There is a need for close monitoring and more diagnostic procedures depending on the detected risks. The intensive prenatal care we did for our patient included monthly prenatal care, screening for urinary and genital infections, frequent Doppler and ultrasound monitoring with biophysical profiling, more frequent blood examinations with biochemistry, urinalysis and repeated culture sensitivity. This kind of specialized care is needed in order to avoid preterm birth, perinatal morbidity and mortality.

A systematic review of cohort studies indicated a specialist clinic and service may prevent preterm birth and improve neonatal outcomes.[4] In a specialized care setting, screening and close monitoring in pregnancy are strategies used to identify complications and provide appropriate treatment and follow-up care. However, the impact of this approach depend on use of effective and timely intervention should problems be detected.[5]

Fetal Genetic Risks

Another concern raised by many women with spina bifida is the fetal genetic risk for congenital malformations, particularly spina bifida. Previous studies report a recurrence risk for NTDs at about 4 to 7% of offsprings of women with spina bifida. However, in a descriptive case series which examined 29 pregnancies

in women with spina bifida, no offspring had spina bifida but about 13% were born with other significant congenital malformations.

While all women in the reproductive age should have sufficient folate supplementation to reduce the risk of having a newborn with spina bifida, recommendations are that women with spina bifida should be taking larger doses of folate than their able-bodied counterparts. A 5-mg daily dose of folic acid for women at high risk of having a child with an NTD has been suggested to prevent it.

In order to screen for neural tube defects, the patient had a congenital anomaly scan done at 20 weeks age of gestation and showed no gross anomaly at the time of the scan. In a diagnostic systematic review by Karim et al., which included thirty studies published between 1991 and 2014, the pooled estimate for the detection of all abnormalities in low-risk or unselected populations was 32.35% (95% CI, 22.45-43.12%). In high-risk populations the detection rate was 61.18% (95% CI, 37.71-82.19%).[6]

Labour and Delivery

To arrive at the decision regarding the mode of delivery, we discussed with our patient the risk and benefits of vaginal and caesarean delivery. We discussed caesarean section and its future pregnancy risks, and its immediate complications like pain, endomyometritis, wound separation/infection, urinary tract infection, gastrointestinal problems, deep venous thrombosis, and septic thrombophlebitis.[7,8] With regards to vaginal delivery, we discussed the process of labor and interventions to make vaginal delivery bearable.[7] The patient was informed about pelvic floor problems associated with vaginal delivery which might complicate her current urinary problems.[9] We also agreed to still consider cesarean delivery if there is dystocia or failure to progress and fetal heart rate tracings that suggest fetal distress.[8] In the previously mentioned case series of 6 pregnant women with spina bifida in Japan, vaginal delivery was done in 4 cases and cesarean section in 2. There were no significant anomalies in the newborns of these patients. The success of pregnancy and delivery was achieved by careful monitoring and management of urological and obstetrical complications.

Considering that our patient has spina bifida, the decision on giving epidural analgesia or systemic anesthesia is also important. Although there is no direct evidence on patients with neural tube defects, a randomized trial of 750 nulliparous women at term, where epidural and systemic anesthesia was compared, the rate of cesarean delivery was not significantly different between the groups (17.8% after intrathecal analgesia vs. 20.7% after systemic analgesia; 95% confidence interval for the difference, -9.0 to 3.0 percentage points; P=0.31). Pain scores after the first intervention were significantly lower after intrathecal analgesia than after systemic analgesia (2 vs. 6 on a 0-to-10 scale, P<0.001). The incidence of one-minute Apgar scores below 7 was significantly higher after systemic analgesia (24.0 percent vs. 16.7 percent, P=0.01).[10]

CONCLUSION

With coordinated multidisciplinary care, women with spinal cord lesions in general have good pregnancy outcomes. With urinary complications being the most common sequela of spina bifida and tethered cord syndrome, more frequent monitoring of clinical and biochemical parameters was warranted to reduce the complications of upper urinary tract infections and preterm labour. In the case of this patient, the multidisciplinary team, under the leadership of the

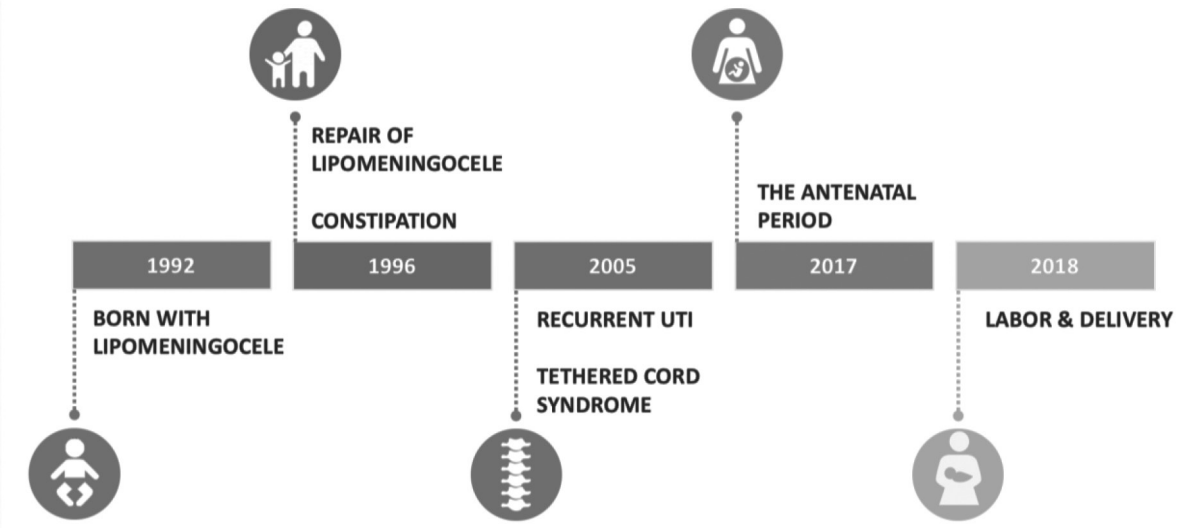

Figure 1. Timeline of patient's symptomatology and course of illness

obstetrician, gave monthly prenatal care, screening for urinary and genital infections, frequent Doppler and ultrasound monitoring with biophysical profiling, more frequent blood biochemistry examinations, urinalysis and repeated culture and sensitivity. Intensive prenatal care is needed in order to avoid preterm birth, perinatal morbidity and mortality.

Genetic predisposition is a risk factor to developing neural tube defects and part of our care is primary prevention for the offspring so that early Folic acid supplementation may be given.

We also gave our patient progesterone throughout the first trimester, although there is no clinical trial on the effectiveness of dydrogesterone among pregnant patients with spina bifida. However, some studies support that it can lower miscarriage rates when given in early pregnancy while the adverse effects were minimal.

With regards to the mode of delivery, careful planning with the multidisciplinary team is advisable, taking into consideration pelvic floor problems associated with vaginal delivery which might complicate her current urinary problems, the probability of dystocia, as well as the most appropriate type of anesthesia to be given. The success of pregnancy and delivery was achieved by careful monitoring and management of urological and obstetrical problems. ■

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