

A Prenatal Diagnosis and Repair of Spina Bifida

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Introduction

A 34 year-old G2P1 woman at 23 weeks gestation was referred to The Fetal Center for abnormal findings on a routine prenatal ultrasound (U/S). Magnetic resonance imaging (MRI) confirmed a single male fetus with spina bifida (SB) extending from the L4 to the S1 level. Figure 1 Additional findings included ventriculomegaly (15 mm in each lateral ventricle), cerebellar herniation to the C2 level (Chiari malformation), and a lemon sign, (concavity of the parietal bones). Figure 2 The family history was negative for SB and the only medication the mother was taking were the recommended prenatal vitamins.

Diagnosis

SB, a congenital defect of the spine in which part of the spinal cord and its meninges are exposed through a gap in the backbone, is typically detected by U/S in the second trimester. A family history of SB, poor maternal nutrition, or an elevated maternal serum alpha-fetal-protein (AFP) may heighten suspicion and prompt further testing.

Treatment

Before the 2011 Management of Meningocele Study (MOMS) was published, the standard of care for SB was surgical closure of the defect shortly after birth. The MOMS trial showed that prenatal repair could limit damage to exposed spinal contents, arrest spinal fluid leakage, reverse hindbrain herniation, and improve fetal outcomes. Repair, usually between 19 and 25 weeks gestation is done by hysterotomy or a laparoscopic procedure [1].

In the MOMS trial, 183 expectant mothers were randomized to either prenatal or postnatal repair. Results showed that prenatal repair reduced the need for shunt placement (40% vs 82%), decreased hindbrain herniation (64% vs 96%), and improved independent walking by 30 months (42% vs 21%). No difference in cognitive scoring between groups was noted [1].

Management

When SB is found on prenatal screening other anomalies such as oral clefts, talipes equinovarus, and cardiovascular or renal malformations and chromosomal abnormalities are important to detect. Amniocentesis for chromosomal microarray can be performed and screening for additional malformations can be done via U/S and/or MRI. Ideally, testing before 20

weeks gestation allows time for further diagnostic or therapeutic procedures.

If prenatal repair is considered, the mother can be referred to a fetal center experienced with SB repair. If the mother meets the inclusion criteria, the maternal and fetal risks and benefits and the expected long-term outcomes are discussed. Table 1 After the procedure, the mother remains in the hospital on tocolytic medications for about a week to prevent preterm labor. Delivery is scheduled via cesarean section at 37 weeks to avoid the risk of uterine rupture. Parents are counseled that the recurrence risk of SB is 1 in 20 in subsequent pregnancies.

After birth, ongoing neurologic, orthopedic, pediatric, and urologic care are important. Neurologically, the signs and symptoms of hydrocephalus, hindbrain herniation, and tethered cord are monitored as the child grows.² Orthopedically, the severity of disability is correlated with the level of the defect. Table 2 However, infants who undergo prenatal repair are three times as likely to have muscle function that is greater than two levels better than expected.¹ Urologically, almost all children with SB have compromised urinary tract function due to sacral innervation of the bladder. Many require clean intermittent catheterization (CIC) and daily anticholinergic medications to decrease incontinence and susceptibility to urinary tract infections. Although the urologic benefits of prenatal repair remain unproven, small studies show a decrease in the need for CIC and medications.^{3,4} Furthermore, because children from the MOMS trial are not yet adults, comparison of sexual function between groups remains unknown.^{5,6} However, paternity rates of 70% have been reported.

Discussion

With an annual incidence of SB at 3.4 per 10,000 live births in the US, almost 4,000 children a year are affected. Although food supplementation with folic acid has decreased the prevalence, other risk factors such as diabetes, obesity, maternal fever, poor nutrition, family history, and certain medications contribute to its development. However, because 95% of cases have no known risk factors, screening of all pregnant women via maternal serum AFP and/or second trimester U/S is crucial as the window of time between detection and the potential for intervention is only a few weeks.

While the MOMS trial documented improved outcomes for infants with SB, discussion of the maternal and fetal risks and benefits are important in the couple's decision-making. Maternal risks associated with prenatal surgery include chorioamniotic membrane separation, oligohydramnios,

placental abruption, spontaneous membrane rupture, uterine dehiscence, and preterm delivery. Chorioamniotic membrane separation occurred in 26% of prenatal repair cases in the MOMS trial (vs 0% in the postnatal repair group) and can lead to the formation of amniotic bands and umbilical cord strangulation.¹ Oligohydramnios occurred in 21% (vs 4%), placental abruption in 6% (vs 0%), and spontaneous membrane rupture in 46% (vs 8%). A third of patients in the prenatal group had areas of uterine dehiscence or scaring at the time of delivery. The average gestational age for the prenatal group was 34.1 weeks, 13% being born before 30 weeks, compared to 37.3 weeks and no deliveries before 30 weeks in the postnatal group. There were no maternal deaths in either group and there was no difference in perinatal mortality rate between groups [1]. Lastly, while the degree of hydrocephalus has improved with prenatal repair, the need for a shunt is directly correlated with ventricle size. Specifically,

patients with ventricles ≥ 15 mm showed no difference in shunt placement [1].

Patient Outcome

Although the MOMs inclusion criteria were stringent, our patient was one of about 25% of referred mothers who met the inclusion criteria for fetal repair. The couple decided to proceed with the surgery and at 25 weeks gestation a five-hour open repair was performed. The mother was hospitalized for one week to monitor for signs of infection and preterm labor before being discharged home. She continued on nifedipine for tocolysis and a scheduled cesarean section was performed at 37 weeks. Both mother and son were discharged home after two weeks.

Maternal	Fetal
Age older than 18 years	SB at T1-S1 level
Singleton pregnancy	Hindbrain herniation confirmed by MRI
BMI (kg/m ²) 40 or less	No kyphosis 30° or greater
Gestational age 19 0/7 – 25 6/7	Normal karyotype or FISH
Cervical length greater than 20mm	US residency
No history of incompetent cervix	No significant anomaly not related to SB
No existing or planned cerclage in the current pregnancy	
No history of uterine anomaly or uterine leiomyomata	
No history of previous hysterotomy in the active segment of the uterus	
Negative HIV and hepatitis B test results	
No history of hepatitis C positivity	
No type 1 diabetes	
No history of Rhesus or Kell alloimmunization	
No history of platelet alloimmunization	
No history of previous spontaneous delivery at less than 27 weeks gestation	
No history of uncontrolled hypertension, chronic hypertension with end-organ disease, or new-onset hypertension in the current pregnancy	
No medical condition that would contraindicate general anesthesia or abdominal surgery	
Placental abruption???	

Hip	flexion	T12, L1, L2, L3
	extension	S1
Knee	flexion	L5, S1
	extension	L2, L3, L4
ankle	dorsiflexion	L4, L5
	plantar flexion	S1, S2
	inversion	L4
	eversion	S1

Table 1 – MRI of spinal defect starting at the L4 level

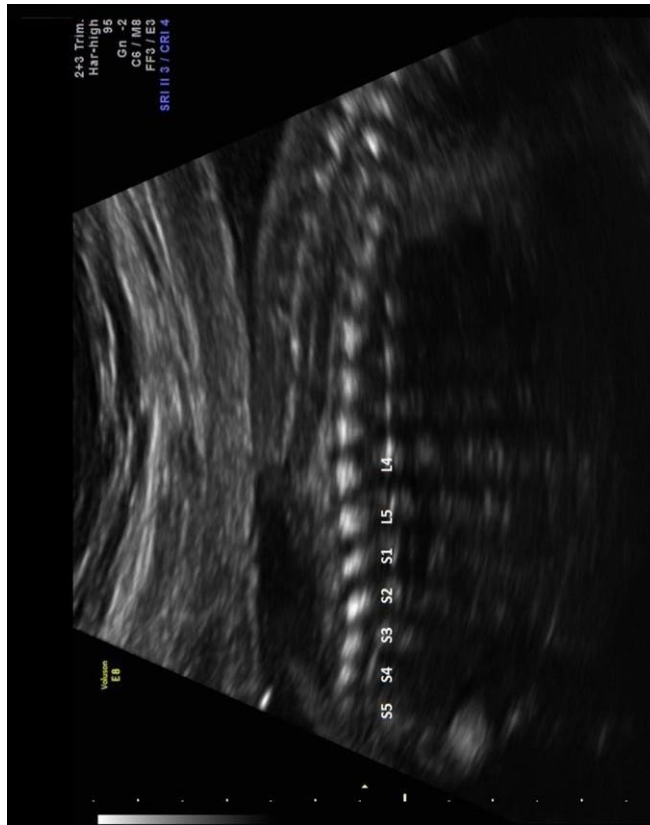


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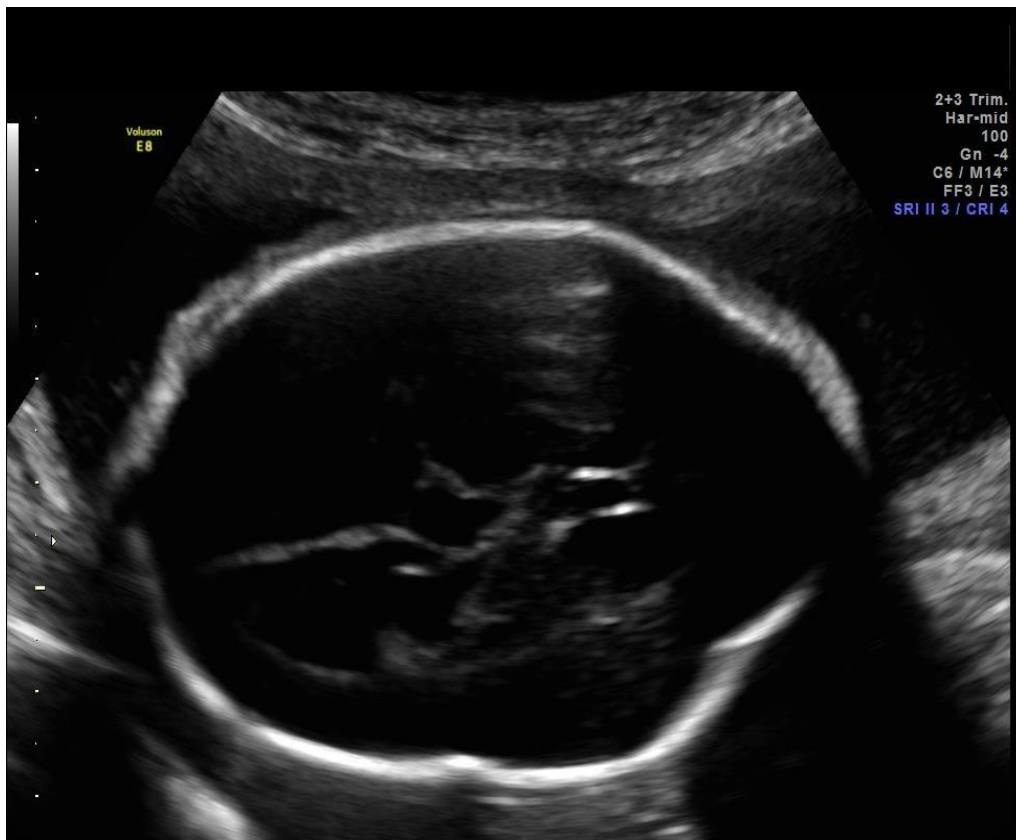


Figure 2 – Lemon-shaped skull

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