Ultrasound Obstet Gynecol 2004; **24**: 595–596
Published online in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/uog.1742

Editorial

Tell the truth about spina bifida

J. P. BRUNER*† and N. TULIPAN‡

*Departments of †Obstetrics and Gynecology and ‡Pediatric Neurosurgery, Vanderbilt University Medical Center, Nashville, TN, USA (*Correspondence. e-mail: joe.bruner@Vanderbilt.Edu)*

Every year in the United States, more than 1000 fetuses in the mid trimester are diagnosed with spina bifida[1]. Many parents have never heard of this malformation, and are therefore dependent upon their local doctors and midwives for initial counseling. Unfortunately, many healthcare professionals are equally ignorant of the current prognosis of children with spina bifida who have ready access to comprehensive care in a modern multidisciplinary clinic. As a result, much of the information initially provided to couples with a newly diagnosed fetus is biased and misleading. As medical ethicists Bliton and Zaner at Vanderbilt University Medical Center observed, 'To date we have met and held intense conversations with more than 150 pregnant women and their partners [diagnosed with spina bifida]. Many times, couples admitted, the initial counseling they received from their obstetrician was slanted – both against disability and toward termination of pregnancy. What they remembered was how the initial obstetric consultation portrayed as grim a picture as possible about their future child's prognosis. Their fears were about mental deficits, ''mental retardation'', paralysis, and their ability to manage the obvious challenges. All were vulnerable to anxiety, guilt, shock, anger, and grief.'[2].

Physicians who routinely tell pregnant women that their fetus with spina bifida will be mentally retarded, never walk, and suffer bladder and bowel incontinence are ignoring a wealth of recent literature that contradicts this stereotype. Most of these children are intelligent, adaptable and able to function well in society. Mirzai *et al.* comment, 'The majority [of children with myelomeningocele] can have a normal IQ and a socially acceptable degree of continence and be able to walk.'[3]. To be sure, newborns with spina bifida possess a spectrum of potentials at birth. The greatest determinant of eventual function, by far, is the upper level of the spinal lesion. In this issue, we discuss prenatal diagnosis of myelomeningocele lesion levels in both community and university hospitals[4]. Armed with this information, healthcare workers and parents alike can access numerous outcome analyses to obtain information specific to the affected fetus.

AMBULATION

Perhaps the greatest concern expressed by prospective parents of a fetus with spina bifida is whether their child will ever walk. In a 25-year follow-up of 71 patients treated at birth in an aggressive, non-selective manner, Bowman *et al.* observed that 46% of young adults (33/71) ambulated the majority of the time (75–100%)[5]. An additional 13% (9/71) ambulated 25–50% of the time. Forty-one percent (29/71) ambulated only with the aid of a wheelchair. When stratified by lesion level, patients with lower defects were more likely to ambulate the majority of the time (Figure 1). Ninety-three percent of patients with a sacral lesion (14/15) ambulated 100% of the time. Ninety-one percent of young adults with an L5 level lesion (10/11) ambulated 75–100% of the time, as did 57% of patients with an L4 lesion. In contrast, no patient with a lesion level of L3 or above ambulated the majority of the time. In a cohort of 73 patients with spina bifida operated on between 1979 and 1993, Mirzai *et al.* reported that almost 60% walked well enough for social purposes[3]. Thirty-seven percent (27/73) ambulated only with wheelchairs and three were bedridden. Finally, among a group of children with spina bifida repaired between 1971 and 1981, Steinbok *et al.* described 53% (44/83) as community ambulators[6]. In brief, approximately two-thirds of children with spina bifida may reasonably expect to ambulate the majority of the time, although almost all ambulators will have lesions at L4 or below. Cochrane and coworkers reported that children they studied participated in hockey, dancing, swimming, basketball, horseback riding, wheelchair sports and competitions, golf and weight lifting[7].

INTELLIGENCE

After walking, the most common concern of parents with a newly diagnosed fetus with spina bifida is intelligence. Bowman *et al.* reported that 85% of the 71 young adults followed by their group were attending or had graduated from high school and/or college, and 63% attended regular classes[5]. Forty-five percent were actively employed, and almost 10% worked as volunteers. Most of the 71 patients lived with their parents, although 11 lived independently and two were married. Mirzai *et al.* noted that 60% of their young adults had normal IQ and development[3]. However, a normal IQ was seen in 76% without hydrocephalus. In McLaurin's series, 70% had a normal IQ[8]. Finally, Steinbok *et al.* observed that 58%

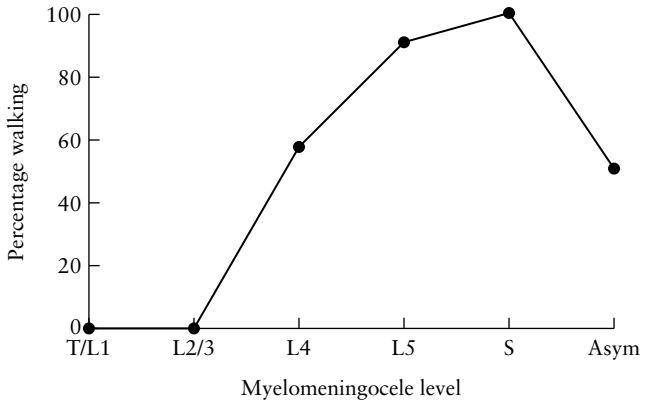

Figure 1 Percentage of study patients ambulating the majority of the time based on myelomeningocele level. (Modified from Bowman *et al.*[5] with permission of S. Karger AG, Basel). Asym, asymmetrical.

of their cohort attended public school, and were in the appropriate grade for their age[6]. Clearly, most children with spina bifida are not 'mentally retarded'.

HYDROCEPHALUS

Historically, almost all newborns with spina bifida received shunts. For example, Steinbok *et al.* reported a 91% shunt rate in 101 children after a minimum follow-up period of 8.6 years[6], a rate almost identical to that of McLaurin[8]. Cochrane studied 85 children with spina bifida and reported that 100% of infants (20/20) with lesions at L3 received shunts, as did 97% (30/31) with lesions at L4–L5, and 91% (31/34) with sacral lesions[7]. Finally, data obtained from the International Myelodysplasia Study Group on 147 patients born between 1980 and 1997 with 'low lumbar' or 'sacral' lesions showed that 94% required shunts[9]. Recent data indicate that these rates may be too high. The Children's Hospital of Philadelphia published shunt rates among 189 children treated for spina bifida between 1983 and 2000[10]. One hundred percent (35/35) of children with thoracic level lesions received shunts, compared with 88% (100/114) of children with lumbar lesions and only 68% (27/40) of children with lesions confined to the sacrum. In recently treated patients, therefore, the upper level of the spina bifida lesion appears to be a major determinant of the need for shunt placement. Fewer neurosurgeons are automatically placing shunts at the same time as the spinal lesion is closed.

CONTINENCE

Although most parents recently informed that their fetus has spina bifida do not express great concern about bladder and bowel incontinence, the issue frequently arises in initial counseling. Bowman *et al.* noted that 85% of their young adults (60/71) used clean intermittent catheterization (CIC), and 90% of those (54/60) performed their own catheterization[5]. For those patients on CIC, 15% always had urinary continence, 68% were dry the majority of the time (75–100%), and 7% were dry 50% of the time. Fifty-two percent of the young adults reported bowel control the majority of the time, and 52% reported 100% social bowel continence. Steinbok *et al.* reported 75% social continence of urine, defined as the ability to remain dry and odor-free without the use of a diaper, and 86% social continence of bowel[6].

THE TRUTH

It should be obvious from this brief review that while spina bifida results in a spectrum of disabilities, with comprehensive medical care most affected children will grow into young adults with normal intelligence, walking, and with social continence of both bladder and bowel. Certainly, a small number of affected children will experience devastating sequelae, but an equal number will be apparently normal, and prenatal imaging is becoming increasingly sophisticated in identifying the extent of disease and likely long-term outcomes. As Bliton and Zaner observed, 'From our discussions with these women and their partners, we came to recognize that ultrasound diagnosis of spina bifida was a momentous event for them that evoked a potentially devastating set of experiences'[2]. Healthcare providers should not allow their own lack of knowledge to magnify their patients' fears of delivering a crippled child. We urge all those involved in prenatal care to learn the facts, and tell the truth about spina bifida.

REFERENCES

1. Centers for Disease Control and Prevention. Folic acid and prevention of spina bifida and anencephaly. *MMWR* 2002; **51**(RR-13): 9–11.
2. Bliton MJ, Zaner RM. Over the cutting edge: How ethics consultation illuminates the complexity of open-uterine fetal repair of spina bifida and patient's decision making. *J Clin Ethics* 2001; **12**: 346–360.
3. Mirzai H, Ersahin Y, Mutluer S, Kayahan A. Outcome of patients with meningomyelocele. *Child's Nerv Syst* 1998; **14**: 120–123.
4. Bruner JP, Tulipan N, Dabrowiak ME, Luker KS, Walters K, Burns P, Reed G. Upper level of the spina bifida defect: how good are we? *Ultrasound Obstet Gynecol* 2004; **24**: 612–617.
5. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA. Spina bifida outcome: A 25-year prospective. *Pediatr Neurosurg* 2001; **34**: 114–120.
6. Steinbok P, Irvine B, Cochrane DD, Irwin BJ. Long-term outcome and complications of children born with meningomyelocele. *Child's Nerv Syst* 1992; **8**: 92–96.
7. Cochrane DD, Wilson RD, Steinbok P, Farquharson DF, Irwin B, Irvine B, Chambers K. Prenatal spinal evaluation and functional outcome of patients born with myelomeningocele: Information for improved prenatal counseling and outcome prediction. *Fetal Diagn Ther* 1996; **11**: 159–168.
8. McLaurin RL. Myelomeningocele: Outcome and late complications. In *Pediatric neurosurgery: Surgery of the developing nervous system*, 2nd edn, Section of Pediatric Neurosurgery of the American Association of Neurological Surgeons (ed). Saunders: Philadelphia, 1989; 53–70.
9. Tulipan N, Bruner JP, Hernanz-Schulman M, Lowe LH, Walsh WF, Nicklaus D, Oakes WJ. Intrauterine myelomeningocele repair: Its effect on CNS structure and function. *Pediatr Neurosurg* 1999; **31**: 183–188.
10. Rintoul N, Sutton LN, Hubbard AM, Cohen B, Melchionni A, Pasquariello PS, Adzick NS. A new look at myelomeningoceles: Functional level, vertebral level, shunting and the implications for fetal intervention. *Pediatrics* 2002; **109**: 409–413.

