## LETTER TO THE EDITOR

## The Prenatal Ultrasonographic Detection of Myelomeningocele in Patients Referring to Children's Hospital Medical Center

## Dear Editor:

Myelomeningocele (MMC) is a major congenital anomaly caused by a defective closure of the neural tube between the 18-25<sup>th</sup> days of gestation.<sup>1</sup> In spite of being a common birth defect with significant lifelong complications, little progress has been made in the postnatal surgical management of the defect and its associated complications.

Over the past decades new screening modalities and diagnostic tests have been introduced that allow earlier and more accurate diagnosis of fetal anomalies, including MMC.<sup>2</sup> As no definite cure is available for MMC, the prenatal detection would help with timely termination of pregnancy as practiced in developed countries.<sup>3</sup>

Studies have shown that the incidence of this malformation is relatively high in Iran, <sup>4,5</sup> and our experience with MMC patients has revealed that most of them are not detected prenatally. Therefore, we retrospectively studied the written prenatal ultrasonographic reports of MMC patients referring to our center to determine the detection rate of MMC.

One hundred forty MMC patients younger than 5 years referring to Children's Hospital Medical Center in Tehran were enrolled into the study from July 2004 to 2005. The median age was 15 days, and 73 (52.1%) were male. The most common sites of MMC were in the following order: lumbar spine in 47 (33.6%), lumbosacral in 40 (28.6%) and thoracolumbar in 25 (17.9%).

All children except 4 (97.1%) had undergone at least one prenatal ultrasound by a sonographist. A first, second and third trimester ultrasound was done in 55 (39.3%), 103 (73.6%) 120 (85.7%) patients respectively. The prenatal sonographic report was normal in 78 (57.4%) patients. MMC and hydrocephalus were both present in only 17 (12.5%). In 16 (11.8%) patients only MMC, and in 25 (18.4%) only hydro-

cephalus was detected. The prenatal ultrasound MMC reports data are shown in Table 1 by the time of detection and level of lesion.

The incidence of MMC in Iran is about 1.6:1000. <sup>4</sup>In developed countries, the incidence of MMC has declined significantly over the past few decades as a result of folic acid supplementation and prenatal detection followed by abortion . The psychological, social and financial burden of the disease on the family and community is enormous, and the care of each affected individual costs about \$250,000 during their lifetime in the United States.<sup>6</sup>

Ultrasound has been reported to be an effective tool for detecting neural tube defects (NTD), and fortunately in Iran it is routinely performed for assessing fetal biometric parameters in most pregnancies. In Iran, there is no national policy on prenatal NTD screening. In the United Kingdom, since the early 1990's, pregnant women have been offered an ultrasound scan at 18-22 weeks.<sup>3</sup> Likewise in France, three ultrasounds are usually performed around the 12<sup>th</sup>, 22<sup>nd</sup> and 32<sup>nd</sup> weeks of gestation. In the USA, there is no such policy regarding the number and timing of prenatal ultrasound studies and they are usually performed on selected patients.<sup>2</sup>

Sonographic screening is mainly based on twodimensional imaging techniques. Doppler and three– dimensional ultrasound scans have not clearly demonstrated their superiority over the conventional two-dimensional ultrasound.<sup>2</sup> The first trimester conventional ultrasound can detect the majority of anencephaly cases, but a significant proportion of abnormalities such as MMC are not detected before weeks 18-20. However, recent reports show that a transvaginal scan in the first trimester for high-risk population can be of great help.<sup>7</sup>

Although MMC is compatible with survival with various degrees of disability, by performing an early prenatal diagnosis during the first trimester, an early pregnancy termination can be made.<sup>3</sup> MMC can be readily screened by the second trimester ultrasound where on the standard "biparietal" cross-sectional view of the fetal lateral ventricles, the "lemon sign" shows the scalloping frontal bones and is predictive of spina bifida. Also, the "banana sign" can refer to the abnormally shaped midbrain and an elongated cerebellum in Arnold-Chiari malformation. Among fetuses with MMC, the "lemon sign" can be detected in 80% and the "banana sign" in 93% of cases. Thus, prenatal finding of an abnormal posterior fossa or ventriculomegaly should prompt the sonographist to rule out an accompanying spinal deformity. Therefore, serial scans of the fetal spine, performed in longitudinal and transversal axes as well as tangential to the skin are needed. An abnormal position of the fetal foot or an enlarged bladder can be the consequence of severe functional impairments, but their absence cannot be considered reassuring .8

In a large study across Europe, MMC detection rate in different countries varied from 33% to 100% with a mean of 68%.<sup>3</sup> Our result with 24.3% overall detection of MMC was far below the international rates, which can be due to a lack of national screening policies for MMC, not adequately looking for MMC on routine ultrasounds and low resolution ultrasound equipment. One of the best solutions to increase MMC detection rate could be referring all pregnant women at weeks 20-22 of gestation to a well equipped ultrasound center with qualified operators for a systematic search for fetal anomalies including MMC. As it is difficult to diagnose MMC in the first trimester, it is recommended that the pregnancy termination time in Iran for this rather prevalent anomaly be increased up to the sixth month of gestation.

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|                           | Diagnosed                 |                           |                           | Undiagnosed | Total     |
|---------------------------|---------------------------|---------------------------|---------------------------|-------------|-----------|
| Level of MMC              | 1 <sup>st</sup> trimester | 2 <sup>nd</sup> trimester | 3 <sup>rd</sup> trimester | n (%)       | n (%)     |
|                           | n (%)                     | n (%)                     | n (%)                     | П (70)      | п (70)    |
| Cervical/Cervicothoracic  | 0 (0)                     | 0 (0)                     | 0 (0)                     | 5 (100)     | 5 (100)   |
| Thoracic/Thoracolumbar    | 2 (6.6)                   | 2 (6.6)                   | 8 (26.8)                  | 18 (60)     | 30 (100)  |
| Lumbar/Lumbosacral/Sacral | 0 (0)                     | 8 (8)                     | 13 (13)                   | 79 (79)     | 100 (100) |
| Total                     | 2 (1.4)                   | 10 (7.4)                  | 21 (15.5)                 | 102 (75.5)  | 135*(100) |

Table 1. Frequency of prenatal ultrasound MMC diagnosis according to the detection time and the level of lesion.

\* One patient was excluded due to double MMC sacs in the cervical and sacral regions that could not be placed in this categorization.