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BRIEF COMMUNICATION

Detection of Open Spina Bifida in the First Trimester: Beyond Intracranial Translucency

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Abstract Intracranial translucency (IT) has been used extensively as an indirect marker to exclude open spina bifida (OSB) in the first trimester. Like any sonographic sign, the use of IT is fraught with issues like non-visualization due to shadowing from bone, poor distinction from the adjacent hypoechoic brainstem, etc. In view of these challenges, researchers have come up with additional markers of OSB in the first trimester. The aim of this paper is to illustrate these sonographic clues so as to improve diagnostic accuracy in cases where obliteration of IT is equivocal.

Keywords Open spina bifida · First trimester · Intracranial translucency · Signs

Introduction

Since its recognition in 2009, obliteration of intracranial translucency (IT) continues to be used extensively as an indirect marker of open spina bifida (OSB) in the first trimester [1]. However, there have been reports of cases of OSB without obliteration of the fourth ventricle [2]. There have been issues like non-visualization of IT due to shadowing from frontal bone, poor contrast differentiation between the hypoechoic brainstem and fourth ventricle as well as misinterpretation of the brainstem or cisterna magna as the fourth ventricle. This has prompted researchers to look for other signs which can provide a clue to the presence of OSB in the first trimester. These signs have been described in the sagittal and axial planes of the fetal head which are evaluated routinely during a first trimester scan. The aim of this communication is to illustrate some of the additional signs of OSB described in the first trimester which can strengthen our diagnostic armamentarium.

Signs in Axial Plane

Aqueduct of Sylvius (AOS): Occiput Distance

Finn et al. [3] observed an abnormal position of the midbrain in the form of its juxtaposition to the occiput as well as a reduction in the distance between the posterior border of AOS and the anterior border of the occiput in fetuses with OSB. They recommended visualization of the midbrain in an axial plane immediately caudal to the plane used for measuring the bi-parietal diameter. Here, AOS is seen like an echogenic box (Fig. 1a). They established a lower limit of AOS-occiput distance of 1.7 mm at a crown-rump length (CRL) of 45 mm and 3.7 mm at 84 mm to detect OSB (Fig. 1b). However, it was felt that this technique required higher degree of operator skill and its routine use in a screening setting was not feasible [4].

Crash Sign

Ushakov et al. [5] described the posterior-caudal displacement of the midbrain and its deformation against the occiput as the crash sign. This is attributed to the leakage of cerebrospinal fluid (csf) from an OSB. This is easily seen in the axial plane in fetus with OSB (Fig. 1c, d).

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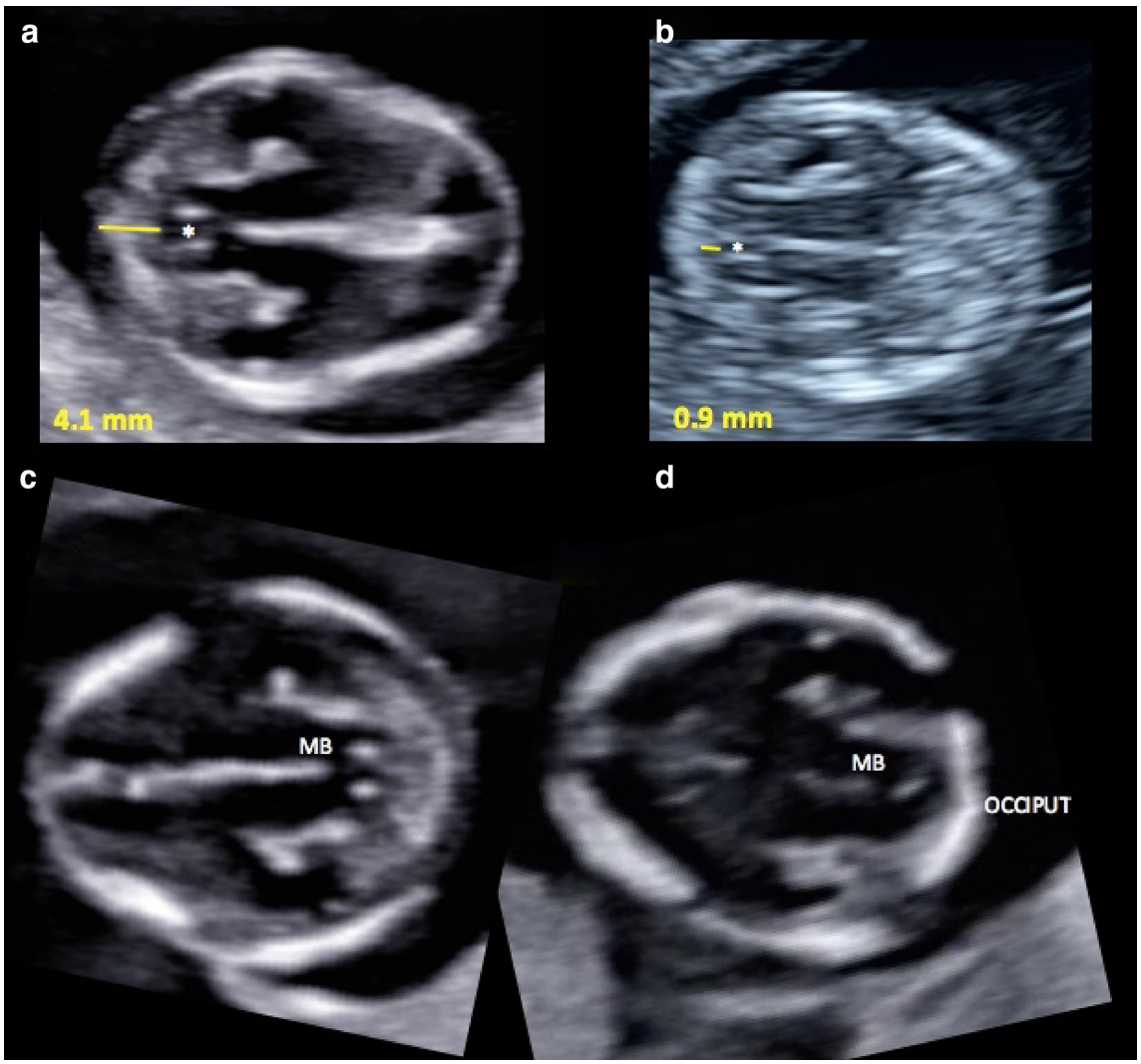

Fig. 1 **a** Aqueduct of Sylvius (AOS)—occiput distance. Axial transabdominal sonogram of head shows normal distance between AOS (*) and occipital bone in a fetus of CRL 68 mm, **b** Aqueduct of Sylvius (AOS)—occiput distance. Axial image shows reduction in AOS—occiput distance in a fetus with myelomeningocele at 12 weeks due to leakage of cerebrospinal fluid. (*) is placed within the csf in AOS. **c** Axial image of head shows the relation of the midbrain (MB) with the occipital bone in a normal fetus at 12 weeks. Normally, the midbrain does not touch the occipital bone. **d** Crash sign. Axial image shows the distorted midbrain (MB) touching the occipital bone in a fetus with open spina bifida (crash sign)

Signs in the Sagittal Plane

Fronto-Maxillary Facial Angle

Lachmann et al. [6] observed a reduction in the fronto-maxillary facial (FMF) angle in fetuses with OSB. The FMF angle is measured in the midsagittal plane between a line along the upper surface of the maxilla and a line drawn from the anterior border of maxilla to the external surface of the forehead. It decreases normally from a mean of 85°–75° with increase in CRL from 45 to 84 mm. However, the FMF angle was less than the 5th centile in 90% of cases with OSB. This decrease is attributed to the caudal displacement of the fetal brain leading to impaired development of the frontal bones. This is also responsible for the lemon sign seen in OSB in the second trimester. Figure 2a, b show the FMF angle in a normal fetus and the reduced angle in a fetus with myelomeningocele respectively.

Maxillo-Occipital Line

Recently, Ramakrishna et al. have described another clue to suspect OSB in the sagittal plane in the first trimester. They drew the maxillo-occipital line along the superior border of the maxilla touching the occipital bone posteriorly [7]. The authors observed that the junction of the thalamus with the midbrain was above this line in normal

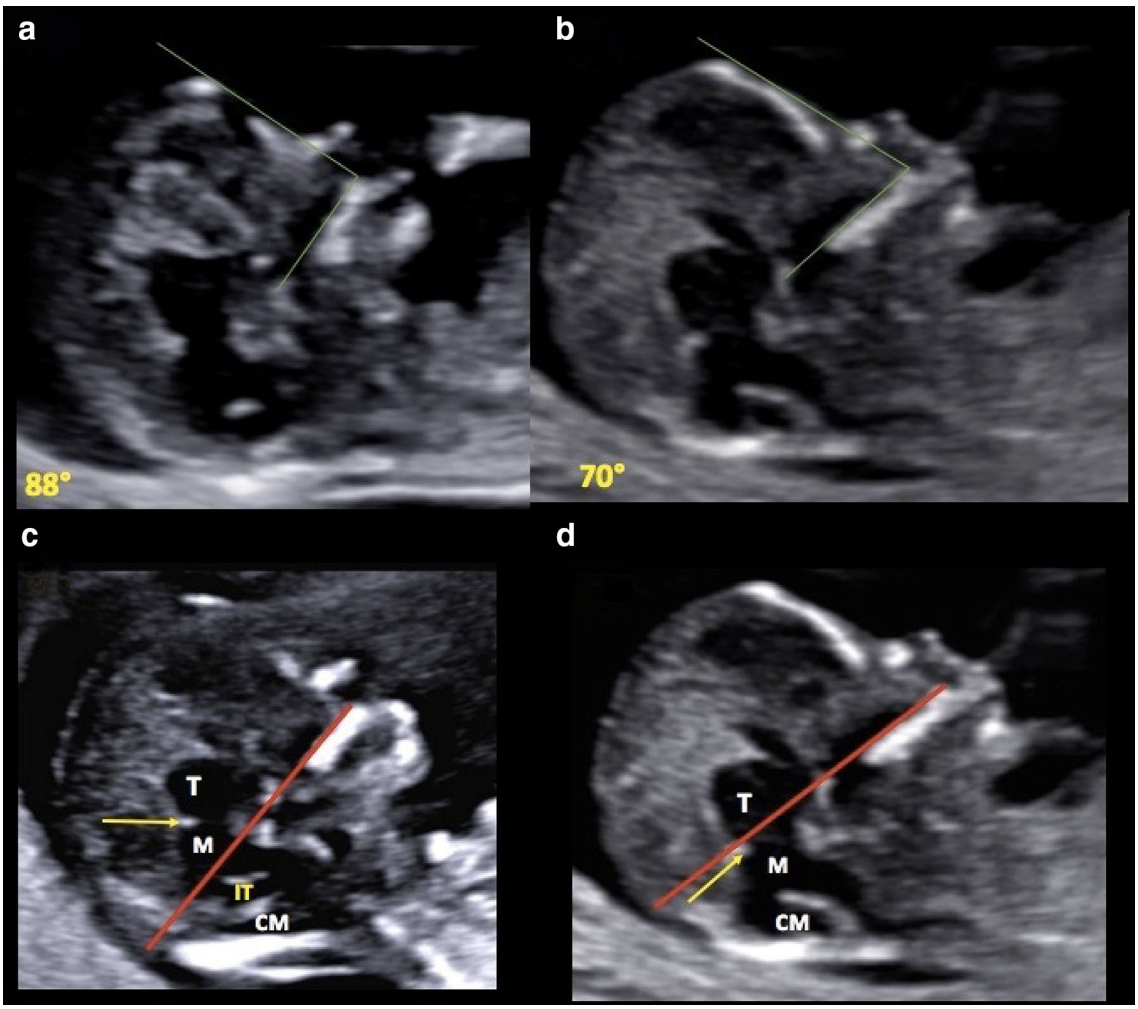

Fig. 2 **a** Fronto-maxillary facial (FMF) angle. Sagittal sonogram of a fetus of CRL 60 mm shows a normal FMF angle of 88°. **b** Decreased fronto-maxillary facial (FMF) angle (70°) in a fetus with open spina bifida at CRL of 68 mm. Obliterated intracranial translucency is also noted. **c** Maxillo-occipital line. Mid-sagittal section of the face in a normal fetus at 12^{+5} weeks. The maxillo-occipital line (red) is drawn along the superior border of the maxilla (MAX) touching the occipital bone posteriorly. The junction (arrow) of the thalamus (T) with the midbrain (M) is above this line. Intracranial translucency (IT); Cisterna magna (CM). **d** Maxillo-occipital line. Mid-sagittal section of the face in a fetus with myelomeningocele at 12^{+4} weeks. The maxillo-occipital line (red) is drawn along the superior border of the maxilla (MAX). The junction (arrow) of the thalamus (T) with the midbrain (M) is below this line. Note the intracranial translucency is obliterated

fetuses and below this line in fetuses with OSB (Fig. 2c, d). This was believed to be due to the descent of the brainstem in OSB owing to the egress of csf.

Conclusion

With the idea of inversion of pyramid of prenatal care, there is a trend towards meticulous first trimester scanning to detect major fetal anomalies. This provides an option of early decision making to the family. OSB is often isolated. At times, there may be only a myeloschisis without any back mass. This makes the direct visualization of the lesion challenging. There have been instances when IT is difficult to interpret due to various reasons. The use of other indirect sonographic markers of OSB can increase our degree of confidence in equivocal cases and facilitate accurate diagnosis.

References

1. Chaoui R, Benoit B, Mitkowska-Wozniak Heling KS, Nicolaides KH. Assessment of intracranial translucency (IT) in the detection of spina bifida at the 11–13-week scan. Ultrasound Obstet Gynecol. 2009;34:249–52.
2. Solt I, Kim MJ, Acuna JG, Adeniji AB, Morales C, Rotmensch S. OC12.02: Spina bifida detection at 11–14 weeks: systematic analysis of technical factors associated with false-positive and

false-negative image interpretation. Ultrasound Obstet Gynecol. 2010;36(S1):22.
3. Finn M, Sutton D, Atkinson S, Ransome K, Sujenthrian P, Ditcham V, Wakefield P, Meagher S. The aqueduct of Sylvius: a sonographic landmark for neural tube defects in the first trimester. Ultrasound Obstet Gynecol. 2011;38:640–5.
4. Chaoui R, Nicolaides KH. Detecting open spina bifida at the 11–13-week scan by assessing intracranial translucency and the posterior brain region: mid-sagittal or axial plane? Ultrasound Obstet Gynecol. 2011;38:609–12.
5. Ushakov F, Everett T, Andreeva E, Tudorache S, Pandya P. P30.04: Crash sign: a marker for spina bifida at 11–13 weeks. Ultrasound Obstet Gynecol. 2016;48:267. https://doi.org/10.1002/uog.16796.
6. Lachmann R, Picciarelli G, Moratalla J, Greene N, Nicolaides KH. Frontomaxillary facial angle in fetuses with spina bifida at 11–13 weeks' gestation. Ultrasound Obstet Gynecol. 2010;36:268–71.
7. Ramkrishna J, Araujo Junior E, White N, da Silva Costa F, Meagher S. P09.11: Maxillo-occipital line: a sonographic marker for the screening of open spina bifida in the first trimester. Ultrasound Obstet Gynecol. 2017;50:184.