

## Case Report

# Fetal ultrasonography of Chiari II malformation

Kumar Sambhav<sup>1\*</sup>, Kumar Dushyant<sup>2</sup>, Shailza R. Jayaswal<sup>3</sup>

<sup>1</sup>Department of Anatomy, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

<sup>2</sup>Department of Anaesthesiology and Critical Care, Max Super Speciality Hospital, New Delhi, India

<sup>3</sup>Department of Radiodiagnosis, Ford Hospital and Research Centre, Patna, Bihar, India

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### \*Correspondence:

Dr. Kumar Sambhav,

E-mail: drkrsambhavaiims@gmail.com

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### ABSTRACT

Chiari II malformation is a complex hindbrain malformation, embryologically presenting as a neural tube closure defect comprising of downward herniation of cerebellum, pons, medulla and fourth ventricle into the spinal canal. We present a case of Chiari II malformation depicting essential sonographic observations at 20 weeks of gestation. Such fetuses have high morbidity and mortality and early diagnosis of such a disease is essential in terms of planning including pregnancy termination or neurosurgical procedures for follow-up pregnancy. Ultrasonography is an indispensable modality for the evaluation of fetal anatomy, growth, and such congenital anomalies.

**Keywords:** Neural tube defect, Spina bifida, Lemon sign, Banana sign, Chiari malformation, Obstetric ultrasonography

### INTRODUCTION

The Chiari malformation was first described by Chiari in 1891, following by Arnold in 1894.<sup>1</sup> Chiari II malformation has an estimated prevalence of 0.44 per 1000 live births and has been observed to be further decreasing owing to prophylactic maternal folate therapy.<sup>2</sup> It is attributed to 3% of all abortions and carries a 1-2% recurrent risk.<sup>3</sup> Chiari malformation is broadly classified into 4 types. Chiari II malformation is observed to be associated with myelomeningocele and hydrocephalus. Essentially, sonographic findings are lemon sign, banana sign, spina bifida and myelomeningocele. We present a case illustrating all crucial ultrasonographic signs for the diagnosis of Chiari II malformation.

### CASE REPORT

A 29-year-old primigravida presented with five-months amenorrhea with no previous history of conception, abortion, chronic illness or any supplementation of folic acid tablets or other prolonged medication. There was no recorded history of neural tube defects in the family. A single live fetus with an estimated gestational age at 20

weeks of age was observed. Anomaly scan was conducted to reveal multiple anomalous sonographic findings of bifrontal indentation or frontal scalloping (lemon sign), banana sign, spina bifida in the posterior vertebral element and myelodysplasia (myelomeningocele) (Figures 1-3). Focal calvarial thinning with shallow posterior fossa was also observed. Foetal growth and amniotic fluid index were noted within normal limits. These characteristic findings lead to confirming the diagnosis of Chiari malformation type II.



**Figure 1: Lemon sign.**



**Figure 2: Banana sign.**



**Figure 3: Myelomeningocele.**

## DISCUSSION

Over the years, studies have documented the accuracy of ultrasonography in the diagnosis of Chiari malformations.<sup>4,5</sup> In the second trimester, a detection rate of 70-90% for foetal congenital abnormalities can be achieved by ultrasonography.<sup>6</sup>

Prenatal sonographies are routinely done for incidental abnormal CNS findings such as Chiari malformation. The documented sonographic findings in Chiari II malformation are lemon sign and banana sign observed as early as 12 weeks whereas myelomeningocele can be identified even earlier at approximately 10 weeks.<sup>7</sup>

Myelomeningocele coexists in 90% of chiari II malformation. Supratentorial abnormalities are the rule in chiari II, not the exception as in chiari I.

Chiari malformations, is currently being described in 4 categories- chiari i : >5 mm descent of the caudal tip of cerebellar tonsils, beyond the foramen magnum; chiari ii: brainstem, fourth ventricle, and >5 mm descent of the caudal tip of cerebellar tonsils past the foramen magnum with spina bifida; chiari iii: herniation of the cerebellum with or without the brainstem through a posterior encephalocele; and chiari IV: cerebellar hypoplasia or

aplasia with normal posterior fossa and no hindbrain herniation.<sup>8</sup>

Characteristically in chiari type 2 malformation, all three structures namely cerebellum, fourth ventricle and medulla oblongata are inadvertently displaced from the foramen magnum towards the cervical canal. Following this there is closure of the cisterna magna and a consequent displacement of the small cerebellum occurs towards the posterior fossa.

Banana sign is the term attributed to this phenomenon.<sup>9</sup> It has also been hypothesised that there is a downward shifting of the brain owing to a decrease in intraspinal pressure with spina bifida. This occurrence is referred to as lemon sign.<sup>10</sup> Both these signs are essential sonographic markers of Chiari II malformation and spina bifida. This lemon sign is readily observed before 24 weeks of gestation, but with further development, it is obscured leading to reduced reliability.<sup>10,11</sup> Moreover, it is also to be borne in mind that lemon sign isn't specific to spina bifida and can be associated with numerous conditions viz. Dandy-Walker malformation, corpus callosum agenesis, diaphragm hernia, and cystic hygroma.<sup>12</sup>

The typical downward herniation of the cerebellum and other structures may lead to non-communicating hydrocephalus essentially by obstructing the flow of cerebrospinal fluid.<sup>8</sup> Hydrocephalus was not evident in our sonogram. The occurrence of myelomeningocele is significant in the context of providing the parents an opportunity to consider and deliberate pregnancy termination. Theories have been put forward regarding the aetiology and causative association between cerebellar herniations and myelomeningocele but have been inconclusive in elucidating the mechanism of this association.<sup>13</sup>

Preconceptual folate therapy has been recommended because of severe morbidity. It has been postulated that nearly half of the neural tube closure anomalies have mutations on the methylene-tetra-hydrofolate reductase gene (MTHFR).<sup>2</sup> Also, maternal folate deficiency and teratogens such as anticonvulsants have been associated with an increased risk of chiari II. In our report, there was no history of anticonvulsant intake nor folic acid intake.<sup>2</sup>

## CONCLUSION

Maternal folate therapy in preconceptional period is vital in the context of high morbidity of neural tube defects. Unplanned pregnancies suffer a greater risk of such defects. Hence, we advocate females to undergo at least 4 weeks of folic acid with vitamin B12 supplementation prior to the time of marriage itself. Neural tube closure happens by about 4<sup>th</sup> week of pregnancy and the preventive effect of folic acid is ineffective after this period. Timely sonographic evaluation of the fetus during antenatal visits is paramount.

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