STM JOURNALS

International Journal of
Midwifery Nursing and Practices

ISSN: 2584-0800
Volume 4, Issue 1, 2026
January–June
DOI (Journal): 10.37591/IJMNP

https://journals.stmjournals.com/ijmnp

Review IJMNP

Lemon Sign: The Diagnostic Indicator for Spina Bifida

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Abstract

The "lemon sign" is a distinctive ultrasonographic finding that serves as a diagnostic indicator for spina bifida, a congenital neural tube defect characterized by incomplete closure of the spinal cord. This sign is observed in fetal imaging and is considered an early and reliable marker for detecting spina bifida, particularly when combined with other prenatal diagnostic tools such as the "banana sign." The lemon sign is characterized by a flattened, concave appearance of the fetal skull, which occurs due to the herniation of the cerebellum and the posterior displacement of the brain. This deformation is a result of the loss of normal intracranial pressure, commonly associated with spina bifida. The identification of the lemon sign during routine fetal ultrasound can significantly aid in early diagnosis, allowing for timely intervention and management decisions. Prenatal diagnosis using ultrasound plays a crucial role in the management of spina bifida, as it enables healthcare providers to offer appropriate counselling, surgical intervention options, and postnatal care planning. This article explores the pathophysiology, clinical significance, and diagnostic accuracy of the lemon sign in detecting spina bifida, providing an overview of how this marker contributes to the diagnosis and subsequent treatment strategies.

Keywords: Lemon sign, spina bifida, prenatal diagnosis, fetal ultrasound, neural tube defect

INTRODUCTION

The lemon sign is a key sonographic marker used in the early detection of spina bifida, especially in high-risk pregnancies, and is most useful before 24 weeks of gestation. It describes a distinctive change in the shape of the fetal skull, as observed during ultrasound. In this condition, the frontal bones lose their normal rounded contour and instead appear flattened or indented inward, giving the skull a lemon-like shape resembling a lemon. This feature is typically identified on transverse ultrasound images of the fetal head taken at the level of the ventricles, where the inward curving (biconcavity) of the frontal bones creates a characteristic appearance [1].

Lemon Sign in Spina Bifida

The "lemon sign" is a radiological feature often observed in fetal ultrasound imaging. It refers to the scalloping or inward concavity of the frontal bones of the skull, resembling a lemon. This sign is strongly associated with neural tube defects, particularly spina bifida and Chiari II malformations.

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Received Date: March 01, 2026
Accepted Date: April 10, 2026
Published Date: April 26, 2026

Citation: Regina P.F. Lemon Sign: The Diagnostic Indicator for Spina Bifida. International Journal of Midwifery Nursing and Practices. 2026; 4(1): 1–6p.

Review of Literature

Studies have consistently emphasized that this sign reflects alterations in fetal cranial morphology, which are often linked to associated intracranial abnormalities [2].

Research findings indicate that the diagnostic value of the lemon sign is enhanced when it is assessed alongside other sonographic indicators, such as the banana sign and ventriculomegaly. The combination of these markers improves the accuracy of early detection and helps identify

associated conditions, such as Chiari II malformation and spina bifida. Evidence also suggests that while the lemon sign is highly prevalent in early gestation, its visibility decreases as pregnancy advances, making early screening crucial to avoid false negatives [3].

Further literature supports the role of advanced imaging techniques, including detailed ultrasonography and fetal MRI, in confirming the findings and evaluating the severity of abnormalities. Early diagnosis using these methods allows for timely decision-making regarding prenatal or postnatal management strategies [4].

In addition, studies have underlined the importance of preventive approaches, particularly adequate folic acid supplementation and early antenatal care, in reducing the incidence of neural tube defects. Broader research also points to ongoing challenges in the detection and management of congenital and rare disorders, especially in resource-limited settings, where awareness, accessibility to diagnostic tools, and cost of care remain significant barriers [5].

Pathophysiology of the Lemon Sign

The lemon sign is closely linked to spina bifida, although its exact cause is not fully understood. One proposed explanation suggests that reduced pressure within the spinal canal in affected fetuses leads to downward displacement of the brain. This shift lowers the intracranial pressure, which in turn affects the shape of the fetal skull, producing a characteristic appearance. As the fetus grows, the frontal bones strengthen and become more resistant to pressure changes, causing the lemon sign to gradually disappear. Additionally, many infants with spina bifida later develop hydrocephalus, and the resulting increase in intracranial pressure may reverse the earlier flattening of the skull [6].

However, this pressure-based theory does not fully explain cases in which the lemon sign is observed despite a normal posterior fossa. An alternative hypothesis proposes that the sign may result from a primary defect in skeletal development, specifically involving abnormal mesenchymal formation of the cranial bone. In some instances of spina bifida, the typical inward scalloping of the frontal bones can be detected as early as the first trimester. This feature may also be related to tethering of the spinal cord at the defect site, leading to progressive downward traction on the brain as the fetus develops [7].

Underlying Cause

The lemon sign occurs due to the abnormal development of the central nervous system in conditions such as spina bifida. Specifically, defects in the spinal cord allow cerebrospinal fluid (CSF) to leak from the spinal canal, thereby reducing intracranial pressure. This leads to a pulling effect on the developing brain and skull, causing the frontal bones to deform inwards.

Association with Chiari II Malformation

The lemon sign is often accompanied by other features of Chiari II malformation, such as cerebellar herniation through the foramen magnum. This herniation further alters the pressure dynamics within the skull.

Transient Nature

The lemon sign is typically observed during the second trimester. In many cases, the appearance of the skull may normalize later in gestation, even though the underlying neural tube defect persists.

Diagnosis and Imaging

- The lemon sign is usually identified during a second-trimester fetal ultrasound (18–22 weeks of gestation).
- It is often accompanied by other ultrasound findings suggestive of spina bifida, including:
  - *Banana sign*: The cerebellum appears compressed and curved, resembling a banana.
  - *Ventriculomegaly*: Enlargement of the ventricles due to obstructed CSF flow.

Clinical Significance

- The lemon sign is an important marker for the early prenatal detection of spina bifida and Chiari II malformations.
- Its presence should prompt further evaluation, including detailed anatomical ultrasound and possibly fetal MRI, to confirm the diagnosis and assess the severity of the condition.

Associated Disease Conditions

- Chiari II malformation
- Spina bifida
- Encephalocele
- Dandy–Walker malformation
- Thanatophoric dysplasia
- Cystic hygroma
- Diaphragmatic hernia
- Corpus callosal agenesis
- Fetal hydronephrosis
- Umbilical vein varix

Radiographic Assessment

The lemon sign is identified on axial images of the fetal head, most commonly during prenatal ultrasound, although it can also be observed on fetal MRI. It is characterized by an inward curving of the frontal bones, rather than simple flattening, giving the skull a distinctive shape [8].

Preventive Measures for Spina Bifida (and Lemon Sign)

Folic Acid Supplementation

The most effective preventive measure for spina bifida is adequate folic acid intake before conception and during early pregnancy.

Recommended Doses

- 400–800 mcg daily for women of childbearing age.
- 4 mg daily for women at high risk (e.g., those with a previous pregnancy affected by spina bifida).

Healthy Maternal Lifestyle

- Avoid smoking, alcohol, and recreational drug use during pregnancy.
- Control conditions, such as diabetes and obesity, are associated with an increased risk of tube defects.

Avoid Certain Medications

Some medications, such as antiepileptic drugs (e.g., valproic acid), are associated with a higher risk of neural tube defects. Women taking such medications should consult their healthcare providers before conception to discuss safer alternatives.

Early Prenatal Care

Regular prenatal visits and early ultrasounds help detect and manage tube defects early in pregnancy.

Treatment Options for Spina Bifida and Related Conditions

Treatment for the lemon sign itself is not required, as it is a marker of spina bifida. However, management focuses on addressing the underlying spina bifida and its complications [9].

Fetal Surgery

Prenatal Myelomeningocele Repair

If spina bifida is detected early, fetal surgery may be performed to close the neural tube defect. This can reduce the severity of Chiari II malformation, improve motor outcomes, and potentially normalize the lemon sign.

Postnatal Surgery

If prenatal surgery is not performed, the neural tube defect is typically repaired soon after birth to prevent infection and further nerve damage.

Shunt Placement for Hydrocephalus

Many children with spina bifida and Chiari II malformation develop hydrocephalus (excess cerebrospinal fluid (CSF) in the brain). A ventriculoperitoneal (VP) shunt or endoscopic third ventriculostomy (ETV) may be required to manage this condition.

Rehabilitation and Supportive Care

Children with spina bifida often require:

- Physical therapy to improve mobility.
- Braces or mobility aids for walking.
- Management of bladder and bowel dysfunction.

Multidisciplinary Care

Spina bifida often requires a team approach involving neurosurgeons, urologists, physical therapists, and developmental specialists to ensure comprehensive care.

Prognosis

- With advancements in fetal surgery and postnatal care, the outcomes of children with spina bifida have significantly improved.
- Early intervention and long-term multidisciplinary care can help maximize mobility, independence, and the quality of life.

The lemon sign is a well-known example of fruit-based terminology used in antenatal imaging. It refers to an inward indentation of the fetal frontal bones, creating a shape that resembles a lemon. This finding is commonly associated with Chiari II malformation and is observed in a large proportion (approximately 90–98%) of fetuses diagnosed with spina bifida.

There is limited data specifically on "Lemon syndrome," as this name might refer to a condition that is not widely recognized or studied under that term. However, India faces significant challenges in managing rare genetic disorders, including conditions such as Lemon syndrome, if they fall within that category.

India has a growing burden of rare diseases, with an estimated 7000-8000 different conditions affecting the population. However, most rare diseases remain undiagnosed because of a lack of awareness, inadequate diagnostic facilities, and high costs associated with genetic testing and treatment. Conditions such as lysosomal storage disorders, metabolic errors, and other inherited disorders are more commonly reported.

Preventive measures for rare diseases in India primarily revolve around early diagnosis through genetic screening, especially in cases of consanguinity, which increases the risk of inheriting genetic disorders. Some strategies include the following:

1. *Genetic counseling*: Offering counseling to high-risk families, particularly those with a history of genetic disorders, to better understand the risks and options.
2. *Prenatal screening:* Advances in genetic testing, including techniques such as whole-exome sequencing, allow for the early detection of disorders before birth.
3. *Awareness and education*: Raising awareness among healthcare professionals and the public can improve early diagnosis and treatment.

India's National Policy for Rare Diseases 2021 aims to provide better access to diagnosis and

affordable treatment of rare diseases. However, implementation and resources remain a challenge due to the high costs of treatment, with many drugs remaining inaccessible to the average patient.

A study involving 1,561 high-risk pregnancies for fetal neural tube defects assessed the frequency and reliability of ultrasound markers, such as the lemon sign, cerebellar abnormalities, head size, and ventriculomegaly. Among the 130 fetuses diagnosed with open spina bifida, the presence of these markers varied according to gestational age. The lemon sign was observed in 98% of cases at or before 24 weeks of gestation but dropped significantly to 13% after 24 weeks of gestation. In contrast, cerebellar abnormalities were consistently detected in 95% of the cases, regardless of gestational age. Before 24 weeks, the most common cerebellar finding was the banana sign (observed in 72% of cases), whereas beyond 24 weeks, cerebellar "absence" became more frequent (81%) [10].

As pregnancy progressed, both growth restriction and ventriculomegaly tended to worsen, whereas the head circumference remained consistently smaller than expected for the gestational age. These findings highlight the importance of early ultrasound diagnosis, particularly before 24 weeks, using indirect markers such as microcephaly, ventriculomegaly, cerebellar changes (notably the banana sign), and the distinctive frontotemporal skull shape known as the lemon sign [11].

CONCLUSION

Detection of the lemon sign during a second-trimester ultrasound should lead to a detailed evaluation of possible neural abnormalities, as it can suggest open spina bifida or other tube defects. However, this sign is not specific and may occasionally be observed in otherwise healthy fetuses. Therefore, it should always be assessed alongside additional imaging findings and clinical information. Studies have shown that the presence of a lemon sign is not related to the level of the spinal defect or the presence of other anomalies. Despite its limitations, it remains a valuable marker, especially in high-risk pregnancies, and plays an important role in enhancing the early ultrasound detection of spina bifida before 24 weeks of gestation.

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