

## Signs in Imaging

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### The Lemon Sign<sup>1</sup>

#### APPEARANCE

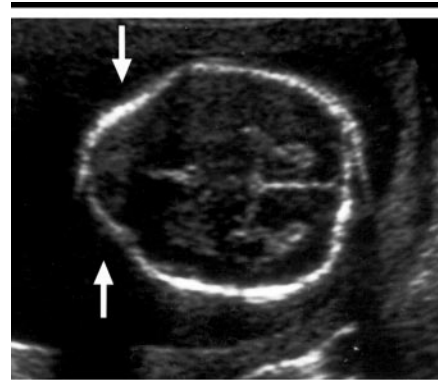
The lemon sign refers to the shape of the fetal skull at ultrasonography (US) when the frontal bones lose their normal convex contour and appear flattened or inwardly scalloped. This gives the skull a shape similar to that of a lemon (Figs 1, 2). The sign is seen on transverse sonograms of the fetal cranium obtained at the level of the ventricles (1,2).

#### EXPLANATION

The lemon sign has a strong association with spina bifida. Although the exact pathogenesis is unknown, it has been postulated that the decrease in the intraspinal pressure in neonates with spina bifida causes the brain to shift downward. This shift decreases the intracranial pressure, which is reflected onto the fetal cranium. The frontal bones are the most vulnerable to the decreased intracranial pressure and respond by flattening or scalloping inward. As the fetus matures, the lemon sign disappears because the frontal bones become stronger and are able to withstand the decreased pressure. In addition, the majority of neonates with spina bifida develop hydrocephalus as they mature. This increase in intracranial pressure can lead to reversal of the flattening (2,3). However, this theory does not explain why the lemon sign is present in fetuses with a normal posterior fossa. Therefore, an alternative theory has been proposed that the lemon sign might be due to a primary skeletal developmental disorder and that the contour of the skull is a result of mesenchymal dysplasia of the cranium (4).

#### DISCUSSION

The lemon sign is very useful in the detection of spina bifida in a high-risk population before 24 weeks of gestation (2). In their



**Figure 1.** Transverse cranial sonogram of a 20-week-old fetus with spina bifida. Image obtained at the level of the ventricles demonstrates the lemonlike configuration of the fetal skull due to biconcavity (arrows) of the frontal bones.

retrospective study of fetuses with open spina bifida, Nicolaidis et al (5) found that the lemon sign was present in all of the 54 fetuses before 24 weeks of gestation; images were obtained at the level of the biparietal diameter. In a subsequent study, Nyberg et al (2) found that in fetuses with spina bifida examined before 24 weeks of gestation, the lemon sign had a high sensitivity (93% [13 of 14 cases]), a high specificity (99% [212 of 215]), and a positive predictive value of 81% (13 of 16). Filly (6) found that when the lemon sign was applied to a low-risk population, the sensitivity and specificity remained high (90% [nine of 10] and 98.6% [9,859 of 9,990], respectively); however, the positive predictive value decreased to 6% (nine of 149), while the negative predictive value remained high at 99.9% (one of 9,9851). As the gestational age of the fetus increases, the lemon sign may disappear and become less reliable in the detection of spina bifida (7).

The lemon sign is not exclusive to spina bifida. It has been seen in a variety of conditions such as encephalocele, Dandy-Walker malformation with encephaloceles, thanatophoric dysplasia, cystic hygroma, diaphragmatic hernia, agenesis of the corpus callosum, fetal hydronephrosis, and umbilical vein varix and two-vessel cord (4). False positive findings have also been caused by fetal triploidy (8). When the frontal contour is only minimally concave or appears linear at US, it is termed the "mild lemon sign." It is important to

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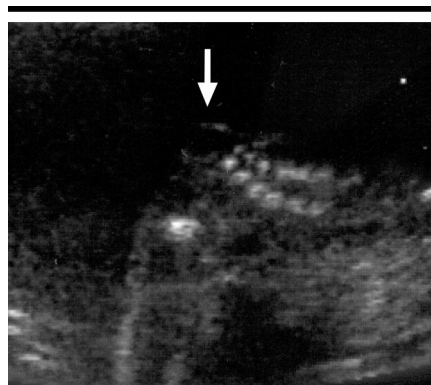


**Figure 2.** Transverse cranial sonogram of an 18-week-old fetus demonstrates the normal contour of a fetal skull.

differentiate this from the true lemon sign, since the mild lemon sign can occasionally be seen in normal fetuses (2). The lemon sign can also be falsely produced by angling the US transducer downward and anteriorly to include part of the orbits (3,9). Regardless of these possibilities, if the lemon sign is present the sonographer should evaluate the spine very closely and also look for other cranial markers of spina bifida (2,9,10). The other fetal cranial markers of spina bifida include ventriculomegaly, microcephaly, obliteration of the cisterna magna with an absent cerebellum, and an abnormal anterior curvature of the cerebellum (3,7).

Neural tube defects are the most common type of severely disabling birth defects in the United States (11,12). Spina bifida affects approximately one in every 1,000–2,000 live births (2). The etiology is multifactorial; both genetic and environmental factors are implicated. Although spina bifida is often an isolated finding, it has been associated with chromosomal abnormalities (trisomies 13 and 18 and partial duplications or deletions of chromosome segments). Factors associated with an increase in the incidence of spina bifida include low parity, low socioeconomic status, relative infertility, diabetes, and obesity. Other risk factors include taking certain medications, particularly anticonvulsants. A woman taking sodium valproate during the first trimester of pregnancy has a 1%–2% chance of having a child with a neural tube defect (3). Folate deficiency in pregnant patients has been associated with spina bifida, and folic acid supplementation has reduced the incidence of spina bifida and other neural tube defects (3,11). Spina bifida occurs most commonly at the lumbosacral region but can occur anywhere along the spine (3). Depending on the level of the spina bifida defect, features vary and include abnormalities or paralysis of the lower extremities, urinary and fecal incontinence, or anesthesia of the skin. Spina bifida is commonly associated with hydrocephalus and Arnold-Chiari II malformations (11).

Prenatal diagnosis of spina bifida is important because early detection before the age of viability allows the patient the choice of a therapeutic termination of pregnancy (2). In addition, there have been recent advancements in fetal surgery that allow repair of the spina bifida defect. The in utero repair may improve the neurologic function of the fetus, as well as improve the hydrocephalus and Arnold-Chiari II malformation (11). Measurement of maternal serum  $\alpha$ -fetoprotein levels is useful in the diagnosis of spina bifida and allows detection of approximately 80% of cases of open spina bifida (2,10). However, it has a low positive predictive value because of the low prevalence of the disease (2). Detection of the spinal defect by



**Figure 3.** Longitudinal sonogram of a fetal spine demonstrates a spinal defect (arrow) covered by membrane.

using prenatal US can be very difficult and depends on the experience and skill of the sonographer. The spinal defect may be detected approximately 80% of the time (16 of 20 cases) when the examination is performed by a highly qualified sonographer who is carefully evaluating the spine (5,10) (Fig 3). In contrast, the sensitivity for detection of a spinal lesion is lower than 50% (five of 14) when US is performed in a low-risk population, by an inexperienced sonographer, or by using less-advanced equipment (2,5). Many facilities have high-risk obstetric departments nearby, and it might be appropriate to refer these patients to the high-risk facilities.

The lemon sign is a useful tool to aid in the detection of spina bifida. Detection of the lemon sign does not require the high level of skill that is needed for US evaluation of the spine. If the lemon sign is present, this should signal the possibility of spina bifida and should prompt the sonographer to look for other cranial markers of spina bifida and to perform a more detailed evaluation of the spine (2,9,10).

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