Signs on Obstetric Ultrasound Images in the Second Trimester

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Abstract
A radiologic sign resembles a specific object, often suggesting a group of similar pathologies. These types of similarities are useful to increase awareness and shorten the list of differential diagnoses. As in other fields of radiology, in obstetric ultrasonography many signs have been described to ease diagnosing. These signs identified on prenatal ultrasonography include the lemon and banana signs observed in spina bifida, teardrop sign in corpus callosum agenesis, keyhole sign in Dandy-Walker syndrome and posterior urethral valve, double bubble sign in duodenal atresia, rabbit lip in cleft lip cases, and in extremity anomalies, frog leg sign in caudal regression syndrome and sandal gap sign. The 2- and 3-dimensional ultrasound images of cases with these signs accompanied by the objects they resemble are presented in this review.

Keywords
Obstetric Ultrasound; Signs; Fetal Anomaly

Özet

Anahtar Kelimeler
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Introduction
In radiology practice some images that are similar to described signs can be useful to classify similar diseases. These signs shorten the list of differential diagnoses and may help doctors reach a final decision for the patient. This report presents the 2- and 3-dimensional ultrasonography images of 9 important and frequently observed radiological signs described on obstetric ultrasound together with sketches and pictures.

Lemon sign
Diagnosis of neural tube defects requires experience and careful examination. In spite of careful examination small open neural tube defects may be missed. As a result cranial anomalies are used as a marker for the presence of spina bifida. A symmetrical flattened lemon sign in the ventral part of the cranium (Fig. 1) shows with neural tube defects and is used as an indicator of spinal opening. The lemon sign is very helpful before 24 weeks gestation in diagnosis of spina bifida in high-risk population; however as gestational age advances as a result of development of the fetus the lemon sign becomes unclear and reliability reduces [1,2]. The lemon sign is not specific to spina bifida. It may accompany encephalocele, Dandy-Walker malformation with encephalocele, cystic hygroma, diaphragmatic hernia, corpus callosum agenesis, fetal hydrops, umbilical vein varices and double-vein cord anomalies. When the lemon sign is present cranial findings such as ventriculomegaly, microcephaly, obliteration of the cisterna magna, compression of the cerebellar hemispheres and ventral-directed orientation (banana sign) should be investigated and the vertebral column requires careful evaluation [3,4].

Banana sign
A sign of Chiari II malformation, as a result of downward migration within the posterior fossa of the fetus the cerebellum wraps tightly around the brain stem and obliterates the cisterna magna (Fig. 2). The cerebellum gains the appearance of a banana. Additionally observed in the majority of spina bifida fetuses, this finding is lost after 24 weeks. This sign may be rarely observed in normal fetuses. Frequently there is accompanying hydrocephaly [5,6]. The incidence in the population is between 0.3-0.7%, and is 2-3% in patients with developmental disorders [7,8]. The etiology varies. Generally it develops due to genetic factors. It shows autosomal dominant, autosomal recessive and X-linked inheritance [9]. Additional anomalies are present in 50% of cases. The most frequent are Dandy-Walker malformation and congenital heart anomalies. Abnormal karyotype (trisomy 18 and 8) are observed at a rate of 20% [10].

Prenatal diagnosis should be suspected in situations of atrium widening and CSP not being observed. Widening of the atrium and occipital horns on axial plane and separation of the body of the lateral ventricle forms a ‘teardrop’ appearance which is specific for diagnosis and frequently found (Fig. 3). As a result of more definite widening at the level of the occipital horn of the lateral ventricles the so-called teardrop configuration (colpocephaly) is observed. The most consistent and identifiable sign is the small appearance of the choroid plexus which appears as a teardrop-shaped pendant (Fig. 3). To confirm CCA coronal or sagittal plane images of cases with dilated atrium and teardrop sign should be taken. Findings such as widening...
of the interhemispheric fissure and third ventricle higher than normal are frequently found. Color Doppler ultrasound showing abnormal branching of the anterior cerebral artery is a supporting finding for diagnosis. In the incomplete type the “tear-drop” sign may be present. However other findings may not be clear. As a result, prenatal diagnosis is difficult. In patients with prenatal diagnosis fetal karyotyping should definitely be completed. While the prognosis is not known for definite, isolated CCA with normal or near-normal development of intelligence has been identified. Some cases are only suspected on US and prenatal MRI is recommended for diagnosis. In the majority of cases diagnosis can be made with MRI [11].

**Keyhole sign (Dandy-Walker malformation)**

Dandy-Walker malformation (DWM) and Dandy-Walker variant (DWMV) are non-specific congenital brain malformations as a result of cerebellum and vermis development anterior-superior or in the rhombencephalon beginning in the ninth week of the embryonic period and not completing by the 16-17th week of pregnancy. It is reported both isolated and as a component of many dysmorphic pathologies. DWM has an incidence of 1/25,000-35,000 in live births. The incidence of Dandy-Walker variant is thought to be higher and it is reported to form one third of posterior fossa lesions. Ultrasonography findings of classic DWM include; larger than normal cisterna magna and/or posterior fossa cyst (cystic dilatation of the fourth ventricle), full or partial agenesis of cerebellar vermis, hydrocephalus or partial expansion of the atriums. In the Dandy-Walker variant ultrasound findings are similar to DWM, though differences include a possibly smaller posterior fossa cyst and varying degrees of agenesis of vermis (full or partial absence of the lower lobe of the vermis). Intracranial ventricular dilatation may or may not accompany this syndrome. The absence of the lower lobe of the vermis creates the “keyhole” observed ultrasonographically which is very useful in DWV diagnosis (Fig. 4) [12,13].

**Keyhole sign (posterior urethral valve)**

Typical presentation of PUV during routine prenatal ultrasound is identification of hydrenephrosis. The large rate of PUV cases identified on prenatal sonography with no evidence identified after birth is very surprising [14,15]. On routine second trimester obstetric ultrasound the PUV anomaly may frequently be accompanied by bilateral hydronephrosis thickening of the bladder wall and in males the keyhole sign at the bladder neck (Fig. 5). This situation forms 10% of all prenatally identified hydrenephrosis [14-16]. The keyhole sign is considered to be very specific to PUV with findings of dilatation of the posterior urethra with posterior urethral obstruction [17].

**Double bubble sign**

In prenatal diagnosis of duodenal atresia, US, karyotype analysis and fetal echocardiography are used. On ultrasonography when the fetal abdomen is shown in the transverse plane the first section of the duodenum, which dilates with the stomach, is full of fluid, showing two separate cystic structures related to each other (double bubble finding) which is used in diagnosis (Fig. 6). This finding is analogous to the gas-filled double-bubble found on radiologic investigation of newborns with duodenal atresia [19]. The double-bubble finding is identified on average at 24 weeks, but it is reported to be identified in earlier weeks [20]. Petrikovsky et al. [21] presented the case of a successful duodenal atresia diagnosis made in the 14th week of pregnancy. In the second trimester, performing a detailed US to identify fetal anomalies in all pregnancies has become a basic application. Parallel to the increase in technology and knowledge more fetal anomalies are able to be diagnosed in the prenatal period. Cleft lip/palate (Fig. 7) is one of the most common congenital anomalies with an incidence of 1 per 1000 live births [22]. In Turkey the incidence of cleft lip/palate is 0.95 per thousand [22]. Cleft lip/palate may be isolated but may also accompany chromosomal, structural anomalies and nearly 350 syndromes [24].
The structures on the midline of the fetal face fully meld by the 7th week of pregnancy. However cleft lip/palate diagnosis may not be possible with high accuracy until the 13-14th week of pregnancy [25]. Recently the use of 3- and 4-dimensional ultrasonography has increased the diagnosis rate of facial defects [26]. Johnson et al. [27] showed the use of 3-dimensional ultrasound had increased the diagnosis rate from 48% to 76%.

**Frog leg sign**

Caudal regression syndrome (CRS) is a rarely-observed congenital anomaly with a variety of degrees of early gestational developmental disorders. This situation is known as sacral agenesis or caudal dysplasia. This malformation is thought to be caused by neuralization defects forming around the 28th day of pregnancy. While maternal uncontrolled diabetes, genetic predisposition and vascular hypoperfusion are possible risk factors the true pathogenesis is unclear. CRS diagnosis is generally given prenatally however again a variable number of newborns with varying degrees of anomalies may be diagnosed. When shorter CRL than expected according to last date of menstrual period and incomplete vertebral ossification on both grayscale and 3-D ultrasonography images are identified, caudal regression syndrome should be considered (Fig. 8). Flexion and immobility of both lower extremities are among the observed findings [28].

Characteristic US findings include sudden interruption to the spine and abnormal positioning of the lower extremities. The femur bones are typically in a fixed V position, appearing like a typical “Buddha pose”. Scanning for possible accompanying urinary and intestinal malformations should be completed [29].

**Sandal gap sign**

The sandal gap deformity is a wider than normal space between the first and second toes and includes medial displacement of the big toe (Fig. 9). The separation of the big toe is reported in 33.3% of Down syndrome cases [30]. However it is not a typical finding for other syndromes. This deformity is observed in many normal fetuses or neonates as a variant of normal. If prenatal ultrasonography shows sandal gap deformity careful scanning for other risk factors of Down syndrome is necessary. If prenatal ultrasonography shows no other abnormalities including clinical risk factors the separation of the big toe should be accepted as a normal variant [31].

**Conclusion**

Described signs with similarity to objects or shapes on obstetric ultrasonography, as in other fields of radiology, carry great importance to ease diagnosis. As a result it is necessary to know the signs and the main pathologies that cause them.

**Competing Interests**

The authors declare that they have no competing interests.

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