

## Sonography of the circumrenal and horseshoe adrenal gland in the newborn

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**Abstract.** The adrenal gland can be visualized using ultrasonography in at least 90% of fetuses of more than 26 weeks gestation. The fetal and neonatal adrenal gland is described as a structure having a characteristic V or Y shape. Despite the frequency of visualization of the neonatal adrenal gland, few reports of adrenal anomalies are present in the radiologic literature. The purpose of this paper is to describe and depict the sonographic appearance of the “horseshoe” and the “circumrenal” adrenal gland and to discuss adrenal anomalies in general.

Adrenal anomalies, usually found in conjunction with renal anomalies, are uncommon, but may be identified using ultrasonography in neonates. To our knowledge, this report is the first to depict the sonographic appearance of the “horseshoe” and the “circumrenal” adrenal gland.

### Case 1

This 3550 g term female newborn was the product of an uncomplicated pregnancy. Although delivery was complicated by transient bradycardia, the Apgar scores at 1 and 5 min were 8 and 9, respectively. Physical examination was unremarkable except for the presence of a single umbilical artery. Renal ultrasound demonstrated that the kidneys were normal in size, shape, and position. The limbs of the left adrenal gland extended below the level of the upper pole of the left kidney and were joined together (Fig. 1). The right adrenal gland had a similar appearance; its limbs extended below the upper pole of the right kidney and joined forming a loop smaller than that of the left adrenal gland. Physical exam was otherwise unremarkable. The neonatal course was uneventful. There was no clinical nor laboratory evidence of adrenal dysfunction.

### Case 2

This term male was the 4280 g product of a pregnancy complicated by polyhydramnios. Amniocentesis was normal. An elective Cesarean section was performed for unstable breech presentation. The

Apgar scores at 1 and 5 min were 5 and 7, respectively. Despite supplemental oxygen the infant remained cyanotic. Physical examination disclosed lowset abnormally shaped ears, a systolic ejection murmur, and a single second heart sound. The infant's cry was soft in amplitude and the child was mildly hypotonic.

Echocardiography on the first day of life demonstrated a left cervical aortic arch, truncus arteriosus, apical muscular ventricular septal defect, a right-to-left shunt at the levels of the foramen ovale and the ventricular septal defect, and a retro-aortic mass. High-resolution real-time ultrasonography disclosed that the retro-aortic mass was a fused (“horseshoe”) adrenal gland lying behind the aorta (Fig. 2). In addition, there was a left pelvic kidney; the right kidney was normal in size, shape, and position. Chromosomal analysis was normal. There was no clinical nor laboratory evidence of adrenal dysfunction.

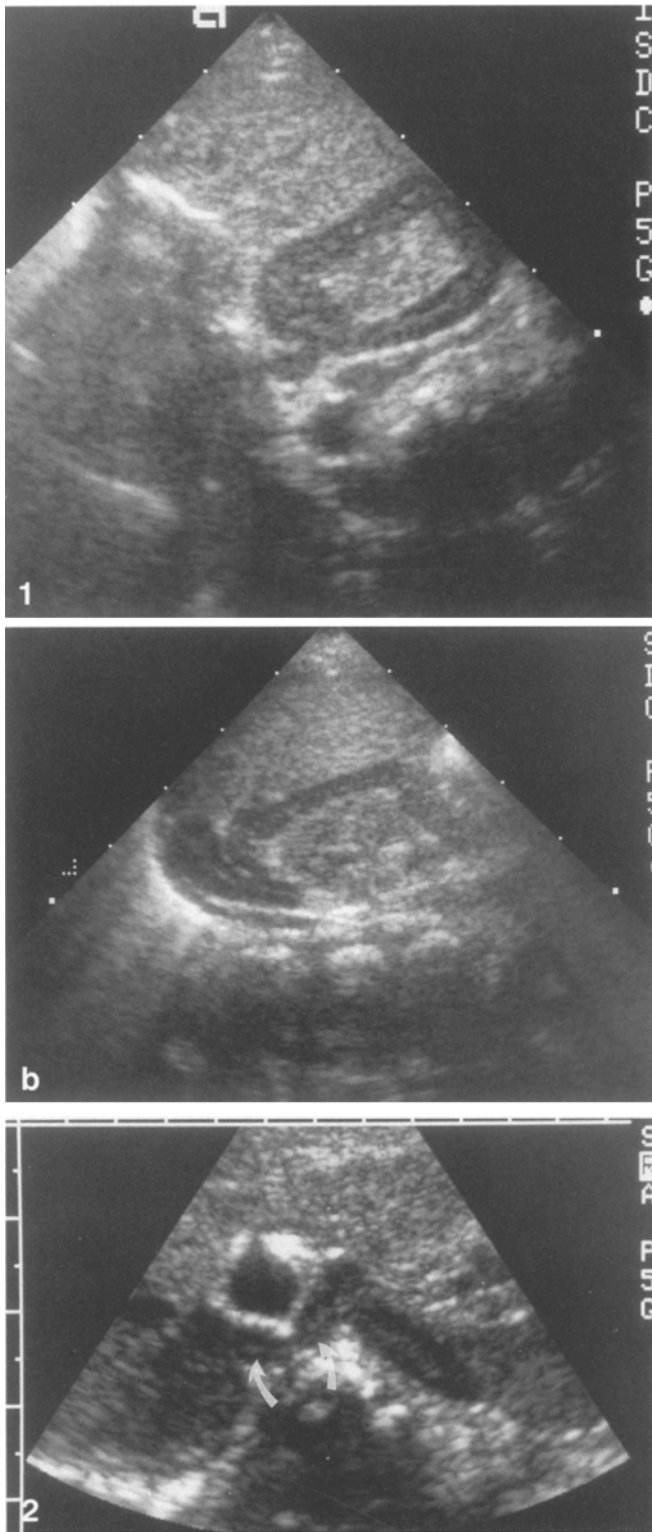
### Discussion

The primitive adrenal cortex is derived from the epithelium of the posterior abdominal wall located between the mesentery and the head of the mesonephros and is visible by the sixth week of gestation. In the third month of gestation, chromaffin cells from the neural crest migrate centrally into the developing adrenal gland and differentiate into the adrenal medulla [1]. Additional proliferation of epithelial cells caps the primitive cortex and forms the outer definitive zone, destined to become the adult cortex [2].

The adrenal gland is relatively large at birth, but by 2 weeks of age the adrenals have regressed by 1/3 of their birth weight. Between 3–12 months of life, the fetal zone is replaced by the zona glomerulosa, fasciculata, and reticularis. By one year of age, the fetal cortical zone involutes and disappears.

By the eighth week of gestation the kidney has migrated cranially to abut the lower pole of the adrenal gland. As the kidney approaches the lower pole, the adrenal gland assumes a conical shape and later becomes pyramidal. Unlike the kidney, the adrenal gland undergoes no marked shift either cranially or caudally during development [1].

In approximately 90% of newborns of more than 26 weeks gestation, the adrenal gland can be visualized



**Fig. 1a,b.** Case 1. Transverse sonograms of the left adrenal gland demonstrate a “circumrenal” contour (a) around the upper pole of the left kidney. The superior limb (1.8 cm in length) of the left adrenal gland is attached (b) and completes the “lasso” appearance. The right adrenal gland (not shown) has a similar appearance, but with a smaller loop to its “lasso”

**Fig. 2.** Case 2. Transverse sonogram demonstrates fusion between the medial limbs of the right and left adrenal glands. The lateral limbs each are 1.7 cm in length. An adrenal isthmus (arrows) posterior to the aorta completes this “horseshoe” adrenal gland

using ultrasonography [2]. Oppenheimer et al demonstrated the right gland in 97% and the left gland in 83% of cases [3]. Typically, the adrenal gland is uniformly thick with a central linear zone of hyperechogenicity and a peripheral zone that is hypoechoic. The central zone is thought to represent the adrenal medulla; the thicker hypoechoic periphery is considered to be the cortex. The majority of adrenal glands in normal neonates are “Y” or “V” shaped; occasionally a “reverse Z,” “omega,” or “trilobed” appearance can be identified [3]. The mean adrenal length is 1.2 cm, 1.4 cm, and 1.7 cm for newborns of 25–30 weeks, 31–35 weeks, and 36–40 weeks gestation, respectively [3]. Using serial ultrasonography, Scott et al noted a decrease of diameter, circumference, and length of the adrenal gland by approximately 50% for each measurement from days 1–42 of life [4].

With the exception of accessory adrenal tissue, congenital adrenal anomalies are considered to be either uncommon or rare [1]. These anomalies include adrenal agenesis, hypoplasia, fusion, or heterotopia. Ageneration of the adrenal gland is thought to be due to failure of the entire nephrogenic ridge, occurring early in the fourth week of gestation. Unilateral agenesis of the adrenal gland is almost always associated with agenesis of the ipsilateral kidney. In 10% of cases of renal agenesis, the adrenal gland is absent on the ipsilateral side [1]. In those instances the adrenal gland is usually flat, discoid, and elongated in appearance instead of pyramidal in shape [5–7]. A similar appearance of the adrenal has been described in patients with renal ectopia [7]. Adrenal hypoplasia usually accompanies anencephaly and may be associated with hypoplasia of the thyroid and gonads, suggesting an abnormal hypothalamic-pituitary relationship in such fetuses [1].

In general, fusion of the adrenals behind the aorta accompanies fusion of the kidney (horseshoe kidney) [1]. Droste et al described one patient with bilateral renal agenesis and a horseshoe adrenal in association with Chiari malformation and meningomyelocele [5]. Case 2 presented here is the first to demonstrate a horseshoe adrenal in association with unilateral renal ectopia.

Adrenal heterotopia may be classified as adrenal ectopia, accessory adrenal tissue, and adrenal rests. In adrenal ectopia, the adrenal gland is in its normal location but lies under the capsule of the kidney or of the liver (adrenal-renal or adrenal-hepatic ectopia). In most cases the heterotopia is bilateral. In these ectopias, a thin connective tissue layer is found between the adrenal parenchyma and adjacent tissue, but this layer is almost always incomplete. Inclusion of the adrenal gland under the capsule of the kidney or liver occurs during the eighth week of fetal development when the ascending kidney comes into contact with a lower pole of the developing adrenal gland [1]. Because the kidney, liver, and adrenal are enveloped in peritoneum in early gestation, fusion between the adrenal and liver or kidney may take place. Similarly, fusion between the limbs of the adrenal may occur if there is subsequent destruction of the intervening layers of coelomic epithelium. The process is analogous to the fusion that may occur between the two kidneys themselves in the formation of a horseshoe kidney [1] and may explain formation of a “horseshoe” or “circumrenal” adrenal.

In conclusion, we have reviewed the embryology and spectrum of adrenal anomalies and have depicted adrenal anomalies in two asymptomatic patients. Because the adrenal glands in the newborn are commonly visualized using ultrasonography, careful attention to the shape and size of the adrenals should allow discrimination of congenital anomalies.

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continued on p. 368