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Prenatal evaluation of the position of the fetal conus medullaris

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KEYWORDS: conus distance; lipomyelomeningocele; spina bifida; spinal cord; tethered cord

ABSTRACT

Objectives *To determine the position of the fetal conus medullaris during pregnancy in relation to the last vertebral body and to examine its use in detecting skin-covered spinal dysraphism.*

Methods *This was a retrospective study involving 300 consecutive ultrasound examinations between 15 weeks of gestation and term. Two operators independently assessed images of the spine to determine whether the conus medullaris and the last vertebral body could be visualized in a single image in a midsagittal plane. The distance between these two landmarks (the conus distance) was measured twice by both operators who were not aware of any previous measurements. Intra- and interobserver variability was assessed by 95% limits of agreement. Linear regression analysis was used to determine the relevant contributors to the conus distance and a normal range was computed based on the best-fit model. The normal results were compared with five cases of prenatally detected skin-covered spinal dysraphism.*

Results *In 84.7% of the 300 cases, both operators were able to visualize the conus medullaris and the last vertebral body. Ninety-five percent limits of agreement for the intraobserver variability in measurement of conus distance were ± 1.9 mm. For the interobserver variability, they were −3.7 and 2.5 mm. We found a linear relationship between conus distance and gestational age, biparietal diameter and abdominal circumference. The strongest relationship was observed for femur length (conus distance = −8.2 + femur length (mm)). In the five abnormal cases, conus distance was well below the 5th percentile.*

Conclusions *Determination of conus distance allows for an objective and feasible assessment of the conus medullaris position. This parameter promises to be useful in the prenatal detection of skin-covered spinal dysraphism. Copyright © 2011 ISUOG. Published by John Wiley & Sons, Ltd.*

INTRODUCTION

Exclusion of spinal defects such as open spina bifida is one of the main objectives in fetal anomaly scanning[1]. In addition to assessment of the fetal cerebellum, examination of the spine involves a careful visualization of the ossification centers and the overlying skin in a transverse, frontal and parasagittal section[2]. In addition to the ossification centers, the spinal cord and the conus medullaris can also be visualized as a dark, triangular structure with two surrounding echogenic lines at the caudal end of the spinal cord. With increasing fetal size, the conus medullaris is shifted towards the fetal head. A low-lying conus medullaris indicates an inappropriate ascent of the spine, which may raise the suspicion of skin-covered spinal dysraphism leading to a tethered cord with or without lipoma, lipomyelomeningocele or diastematomyelia. Although the prevalence of these defects is estimated to be 2–4 in 1000 in postnatal life[3,4], these disorders are rarely diagnosed prenatally[5]. However, early detection is not only important to inform and prepare parents for the anomaly but also to allow pediatric neurosurgeons to develop ahead of time a well-timed surgical repair strategy to avoid irreversible neurological damage[6].

Appropriate screening for these defects requires detailed knowledge about the normal position of the conus medullaris during fetal life. Nevertheless, only a small number of studies describe the level of the conus prenatally[7–10]. In these studies, ascent of the conus medullaris was determined using vertebrae as anatomical landmarks. However, studies comparing the prenatally estimated level of open spina bifida with postnatal results have demonstrated that this assessment is often prone to errors[11].

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In this study, we examined whether the position of the conus medullaris can be assessed by measuring the distance between the conus medullaris and the last vertebral body and whether this measurement can help to detect skin-covered spinal dysraphism leading to a tethered cord.

METHODS

At the Department of Prenatal Medicine of the University of Tuebingen, Germany, the fetal spine is assessed during each ultrasound examination to exclude open spinal dysraphism. As part of this examination, the spinal cord and the conus medullaris are routinely visualized. The integrity of the fetal spine is documented by ultrasound images showing the cerebellum and the fetal spine in a transverse, sagittal and frontal view.

In addition, during each examination, we try to visualize the spinal cord, the conus medullaris and the vertebral bodies up to the end of the spine in a single image to exclude spinal defects other than open spina bifida (skin-covered spinal dysraphism) (Figure 1). The protocol for correct image acquisition involves visualization of the conus medullaris as a distal part of the spinal cord in a midsagittal plane. The conus medullaris can be identified as a dark triangular structure with two surrounding echogenic lines at the caudal end of the spinal cord. In addition, the transverse ossification center of the last vertebral body should be displayed. In the second trimester, at least three sacral ossification points should be visualized; in third trimester at least four should be visualized.

A retrospective search of our digital database was done to select all images of the conus medullaris from 300 consecutive pregnancies between 15 weeks of gestation and term that resulted in healthy live births. Between the 3rd and 10th days following delivery, every neonate underwent a neurologic examination by a pediatrician to exclude spinal defects. Pregnancies were included only once in the study. Cases where it was not possible to identify the conus medullaris and the last vertebral body were excluded from further analysis.

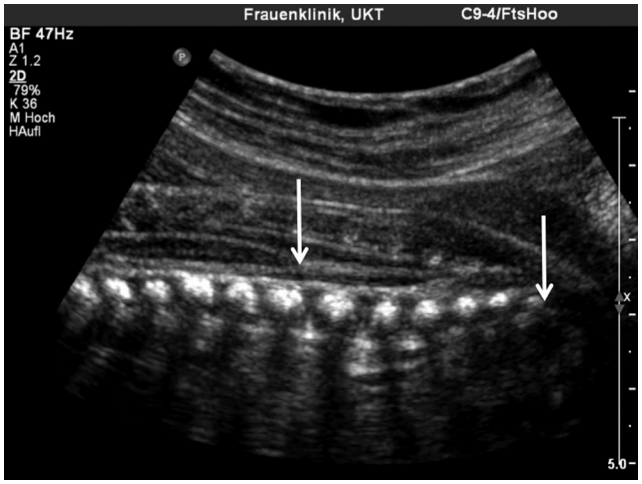

Figure 1 Measurement of the conus medullaris. The triangular conus medullaris is identified in the midsagittal plane. Its distance (between arrows) to the last ossification center of the spine is measured in mm.

The images were assessed by two operators to determine whether they correctly showed the conus medullaris and the last ossification center of the vertebral body. In addition, the distance between the most caudal point of the conus medullaris and the last ventral ossification of the os sacrum (conus distance) was measured (Figure 1). Numeric displays on the screen were covered, so that the operators were blinded to the actual measurements and unaware of previous results. In addition to this first study group, we included five cases of prenatally identified skin-covered spinal dysraphism and compared the conus distance in both groups.

Statistical analysis

The conus distance was measured by two operators. Interobserver reliability was assessed by Bland–Altman plots with 95% limits of agreement (LOA). For intraobserver reliability, the first 30 measurements were repeated by both operators and compared by 95% LOA. For the further analysis, for each case, the mean value of the measurements of Operator A and Operator B was used. Linear regression analysis was used to determine the significant covariates for the conus distance and to produce a model for the normal range. In the abnormal cases, the conus distance was also measured by the two operators and the mean value was compared with the normal range of the normal cases.

RESULTS

In 254 (84.7%) of the 300 cases, the conus medullaris and the last vertebral body were seen and both operators were able to successfully measure the conus distance. The remaining 46 cases were excluded from further analysis. Median gestational age at the time of the ultrasound examination was 23.6 (range, 15.0–40.9) weeks of gestation, mean maternal age was 32.0 (range, 14.9–44.6) years and mean maternal weight was 73.3 (range, 50.9–127.8) kg.

Intra- and interobserver reliability

Mean difference in conus distance between the 30 repeated measurements of Operators A and B was 0.0 mm (95% LOA, −1.9 to 1.9 mm). Mean difference between the 254 measurements of Operators A and B was −0.6 mm (95% LOA, −3.7 to 2.5 mm). The Bland–Altman plot is shown in Figure 2.

Normal range

Mean conus distance was 38.4 ± 15.0 (range, 7.7–76.7) mm. Linear regression analysis showed a significant association between the conus distance and gestational age ($r = 0.948$, $P < 0.001$), femur length ($r = 0.954$, $P < 0.001$), head circumference ($r = 0.953$, $P < 0.001$) and

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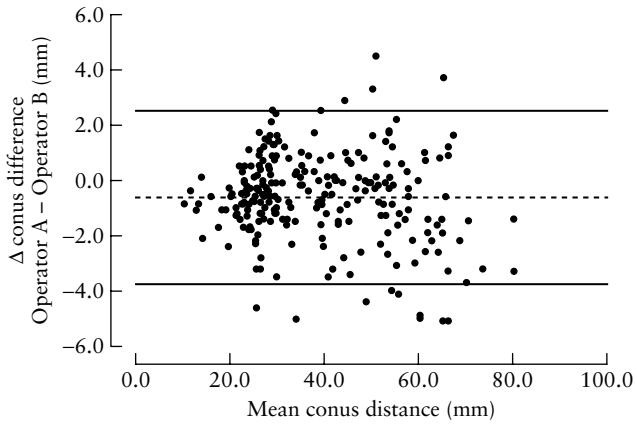

Figure 2 Bland–Altman plot showing interobserver variability in measurement of conus distance between the two operators.

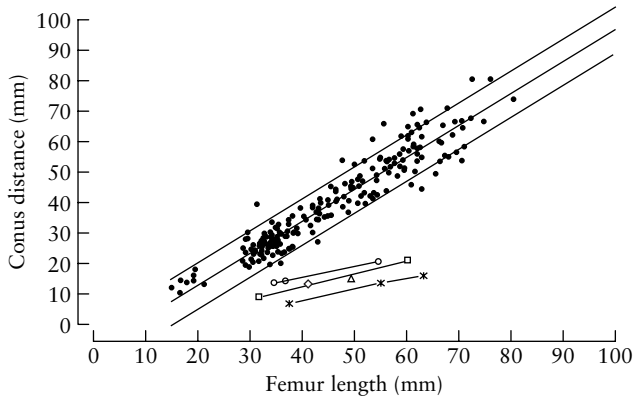

Figure 3 Linear correlation between conus distance and femur length in normal cases and in five abnormal cases (○,◇,△,□,✳). Serial measurements in the five pathological cases are shown.

abdominal circumference ($r = 0.944$, $P < 0.001$). The best-fit formula, between conus distance and femur length, was conus distance $= -8.2 + \text{FL}$ (mm) (SD, 4.5 mm). Figure 3 shows the normal range together with the 5th and 95th percentiles.

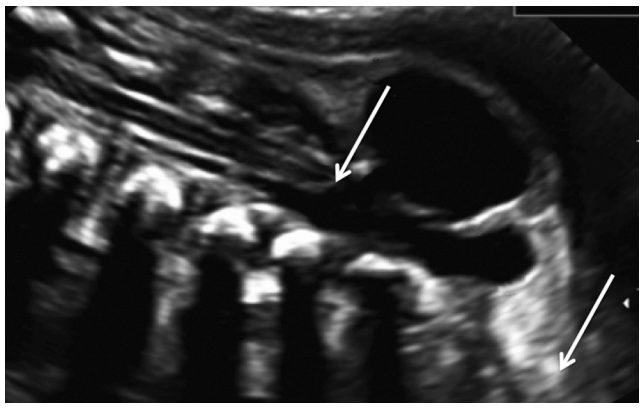

Figure 4 Case with prenatally diagnosed lipomeningocystocele (24 + 4 weeks of gestation). Conus distance (between arrows) is shortened. The conus was not involved in the subcutaneous tumor.

Conus distance in skin-covered spinal dysraphism

Table 1 summarizes the postnatal abnormalities in five prenatally identified cases of skin-covered spinal dysraphism. There were three cases with lipomyelomeningocele. Two cases showed a tethered cord without lipoma. In one case, the fetus demonstrated diastematomyelia with syringomyelia. In the other fetus, the tethered cord was part of a caudal regression syndrome with partial agenesis of the os sacrum. Figures 4 and 5 show ultrasound images of the conus distance indicating the inadequate ascent of the spinal cord. In those cases, mean conus distance was 14.4 ± 4.1 mm, which was well below the 5th percentile. The corresponding z-scores ranged from −3.2 to −6.3.

DISCUSSION

In this study, we have shown that in most cases, a routine assessment of the conus medullaris is feasible and measurement of the conus distance may help to detect skin-covered spinal dysraphism featuring a tethered cord. We found a linear relationship between conus distance and gestational age, biparietal diameter and abdominal circumference, but the strongest relationship was observed with femur length. The regression formula is easy to use, as the expected conus distance can be calculated as

Table 1 Summary of five cases with skin-covered spinal dysraphism

GA at first diagnosis (weeks)	*Absolute conus distance in mm (z-score)*	*Prenatal diagnosis*	*Postnatal diagnosis*	*Additional anomalies*
20 + 3	8.9 (−3.6)	Skin-covered spina bifida	Lipomyelomeningocele	
21 + 1	13.8 (−3.2)	Skin-covered spina bifida	Lipomyelomeningocele	Anal atresia
24 + 4	13.3 (−4.8)	Lipomeningocystocele	Lipomyelomeningocele	
21 + 0	7.1 (−5.3)	Spinal lipoma with tethered cord	Diastematomyelia with syringomyelia	
27 + 2	15.1 (−6.3)	Partial agenesis of os sacrum and tethered cord	Partial agenesis of os sacrum + tethered cord	Structural chromosomal aberration (deletion 7q), unilateral renal agenesis, microcephaly, partial agenesis of corpus callosum

GA, gestational age.

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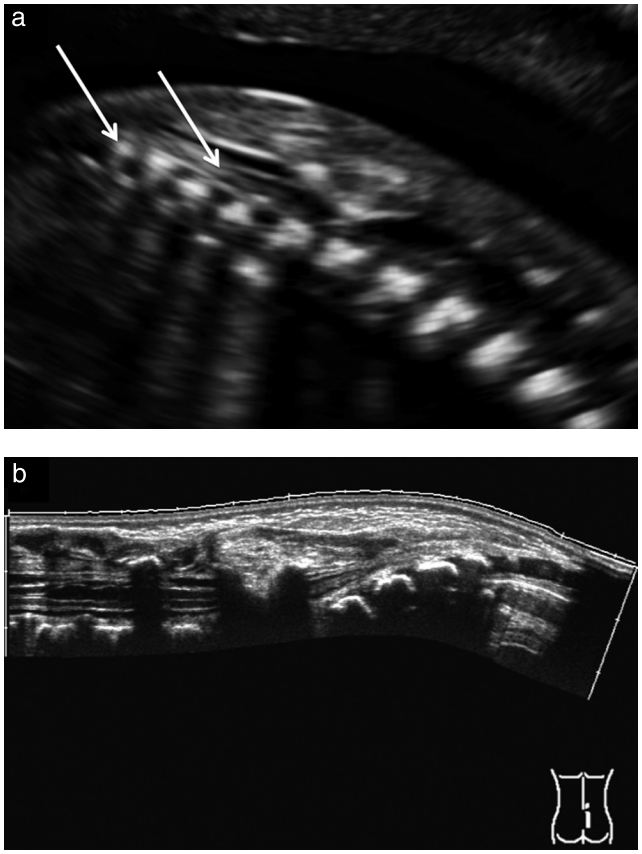

Figure 5 Case with skin-covered spinal dysraphism without visible tumor at 21 + 0 weeks of gestation: subcutaneous lipoma and tethered cord in a case of diastematomyelia. a) Prenatal image at 21 + 0 weeks of gestation showing the shortened conus distance (between arrows); b) postnatal sonographic image demonstrating the diastematomyelia with tethered cord and subcutaneous lipoma.

femur length minus 8. In the five abnormal cases, conus distance was well below the 5th percentile of the normal distribution curve.

There is an ongoing debate about the position and the ascent of the conus medullaris in fetal and neonatal life. Even in postnatal studies, the normal position and the ascent of the conus medullaris remain a matter of debate. Some studies found that the conus ascends to L1, which is considered to be the final position, between 2 and 12 months after birth[12,13]. In contrast, in an autopsy study involving 115 fetuses, it was shown that the conus reaches its final position at birth[14]. Beek *et al.*[15] examined premature infants and found that already at 34 weeks of gestation, the ascent is finalized.

The results of prenatal studies are also confusing. Robbin *et al.*[7] assessed the position of the conus medullaris at 19 weeks' gestation and found that it was at about L2/3, and in some cases, already at about L1/2. Zalel *et al.*[9] examined the conus in 110 fetuses between 14 and 40 weeks' gestation. They found, that in all fetuses between 13 and 18 weeks' gestation, the conus was at L4 or lower. In 97% of the cases, they observed a shift towards L2/3 between 18 and 24 weeks. This was recently confirmed by Perlitz *et al.*[10], who found that at 20–24 weeks of gestation, the conus is between L2 and L3 in 93% of cases.

The reason why the results of these pre- and postnatal studies are so divergent remains unclear. It can be speculated that the correct classification of the vertebral body may be more difficult than expected either due to the position of the fetus, to the curvature of the spine, or to the fact that clear landmarks for orientation are missing around the lower part of the spine. In fetuses with open spina bifida, Kollias *et al.*[11] demonstrated that there was disagreement between the ultrasound and pathological classifications of the height of the spinal defect in 36% of cases. Similarly, in a postmortem MRI study by Widjaja *et al.*[16], two operators were asked to assess the position of the conus medullaris and disagreed in more than half of the cases. Therefore, the conus distance seems to be a better way to describe the position of the conus medullaris, as it appears to be feasible in most cases with low inter- and intraoperator variability.

The rationale for measuring the conus distance relies on the attempt to detect skin-covered spinal dysraphisms such as lipomyelomeningocele, diastematomyelia or other cases of tethered cord. In these conditions, there is an abnormal traction on the conus, the roots of the lower lumbar nerves and the whole spinal cord. Affected children may present with motor and sensory deficits in the lower limbs, abnormal reflexes, neurogenic foot deformities, scoliosis, and bladder and/or bowel dysfunction. In general, these symptoms develop during the first years of life, but in some cases they can be present even at birth. Early recognition and surgical release of the tethered cord is crucial to preserve normal function, prevent deterioration of existing deficits or even induce gradual neurological improvement. Cases of tethered cord deficits acquired after birth may be irreversible if treatment is delayed[6,17,18]. Therefore, prenatal diagnosis of skin-covered spinal dysraphism is of great value for parents, neonatologists, pediatric neurologists and neurosurgeons, and can improve the timing of postnatal diagnosis and treatment. So far, prenatal or even neonatal diagnosis in the absence of a lipomyelomeningocele presenting as an obvious tumor has been rarely described[4,19,20].

In conclusion, measurement of the conus distance is a feasible way to assess the position of the conus medullaris. This may improve the detection of skin-covered spinal dysraphisms.

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