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# Primary Acalvaria in a Chihuahua Dog

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**Abstract :** A three-week old female Chihuahua dog weighing 80 g was presented with the absence of skull palpation, suckling disability, and growth retardation. Physical examination revealed asymmetry of the lips slanting to the left side when feeding milk. Also there were head tilt to the left side and corneal ulcer of the left eye. The absence of the frontal and parietal bones were shown on radiographs. Lateral ventricular enlargement was identified on the ultrasonographic examination. On computed tomographs, frontal and parietal bone defect, ventricle enlargement, and intracranial arachnoid cyst were observed. The dog was dead after 1 day of presentation. The dog was diagnosed as the primary acalvaria by necropsy.

***Key words*** **:** acalvaria, skull defect, CT, US, dog.

## Introduction

Primary acalvaria is a rare congenital anomaly in which the flat bones of the cranium, dura mater, and associated muscles are absent(3,4,7,8). The underlying brain structures are generally complete but have been shown being abnormally developed(3).

The pathogenesis of acalvaria is obscure, however the current belief is faulty migration of mesenchyme(3,4). Migration of the membranous neurocranium does not occur, resulting in the absence of calvaria and dura(4).

Diagnosing skull defects can be evaluated by imaging techniques, such as radiographs, ultrasonographs(US), computed tomographs(CT)(1,7,9). Criteria for diagnosis of acalvaria include absence of calvarial bones with presence of cerebral hemispheres(4,13). Additionally, it is possible to distinguish primary neural tube defects from secondary neural tube defects by the presence of a skin layer overlying the brain matter(3).

To our knowledge, there have been several reports for describing neural tube defects and congenital malformations of the central nervous system in veterinary medicine. Acalvaria, however, has not been reported associated with neural tube defects in veterinary medicine. This case report describes the imaging characteristics of primary acalvaria in a Chihuahua dog.

## Case

A three-week old female Chihuahua dog was presented with the absence of skull palpation, suckling disability, and growth retardation comparing to the other littermates. The dog was only weighing 80 g.

Calvarium was not palpated on the physical examination. Also it revealed asymmetry of the lips slanting to the left side, when feeding milk. There were hypertelorism, head tilt to the left side, and corneal ulcer of the left eye. The dog was positioned herself as left lateral recumbency.

On radiography(ASG-525RF®, Asia X-ray, Korea), loss of frontal, parietal, and occipital bones were shown. The cranium shaped as dome-like appearance and showed homogenous opacity(Fig 1). No other abnormalities were evaluated on thorax, abdomen, and limbs. Through calvarial defect

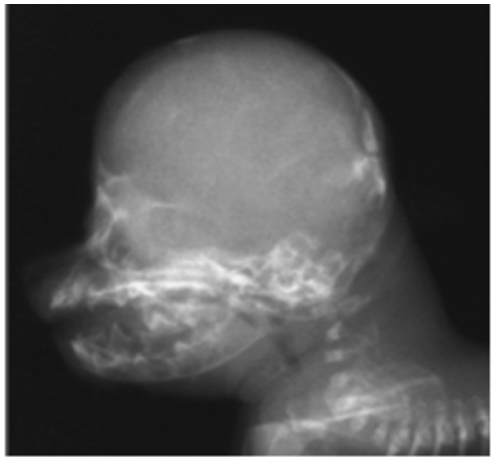

**Fig 1.** Right lateral skull radiography of the dog. Loss of frontal, parietal, and occipital bones is shown. Only some part of the occipital bones is observed (arrow head). The calvarium had dome-shaped and homogenous opacity.

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lesion, the US(SonoAce8800®, Medison, Korea) showed asymmetrical lateral ventricular enlargement. While base of the skull was appeared with acoustic shadowing and disorganization of cerebral parenchyma was not appeared(Fig 2). CT(CTMax640®, GE, USA) was preformed for more detailed and definitive evaluation of the skull and intracranial contents. The bone window CT images showed loss of frontal, parietal, and part of occipital bones(Fig 3). Asymmetrical ventricular enlargement was appeared on the soft tissue window CT images(Fig 3). In addition, intracranial arachnoid cyst(ICAC) was also shown on cerebellar tentorium region(Fig 3). The dog was dead after 1 day of hospitalization.

General necropsy documented dome-shaped brain and the abnormal cranium bones. The loss of frontal, parietal, some part of occipital bones, muscles, and meninges was observed. Therefore the diagnosis was confirmed as primary acalvaria concurred with hydrocephalus and ICAC.

## Discussion

Acalvaria is usually associated with a closed neural tube defects, such as hydrocephalus and holoprosencephaly(7). Neural tube defects, which are the neurologic disorder from developmental abnormality, can be classified as open, if neural tissue is exposed or covered only by membrane, or as closed, if the defect is covered by normal skin(8).

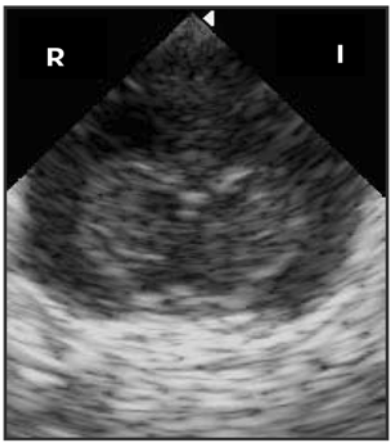


**Fig 2.** Transverse sonogram of the brain in the dog. Anechoic asymmetrical ventricular enlargement is shown.

In postneurulation defect or closed neural tube defect, the embryonic ectoderm is normally situated, such that the ensuing defects are separated from the amniotic fluid by a skin covering(4). This skin covering becomes essential to protect the developing brain, which is thought to be destroyed in anencephaly by continued exposure to amniotic fluid(4). However, in acalvaria, migration of the membranous neurocranium apparently does not occur, resulting in the absence of calvaria and dura with normal skin covering(4). The base of the skull, which is derived from the cartilaginous portion of the neurocranium, develops normally, as does the viscerocranium, the progenitor of the facial bones(4). The pathogenesis of acalvaria is not exactly known(8). Some theories of development of acalvaria suggest that it result because of the primary non-closure of the neural tube or may be a part of a spectrum of anencephaly(8).

Epidemiological survey demonstrates a female predilection in acalvaria in human medicine: gender was reported in 15 of the cases, and 10 of the fetuses were female(4). The condition is not believed to have a specific risk of recurrence. There have been no chromosomal abnormalities in cases of acalvaria(4).

Some congenital anomalies are associated with acalvaria(3,4). Occurrence of other congenital anomalies is common(4). These include holoprosencephaly, hydrocephalus, micropolygyria, hypertelorism, omphalocele, cleft palate and lips(3,4,8). When these anomalies are present with skull defect, acalvaria should be considered.

Diagnosis of acalvaria is based on the diagnostic imaging modalities, such as radiography, US and CT. In human medicine, especially transvaginal US is the best modality for diagnosing acalvaria of fetus. A radiography of the skull demonstrates absence of the calvarium bones, especially membranous neurocranium, such as frontal, parietal, or

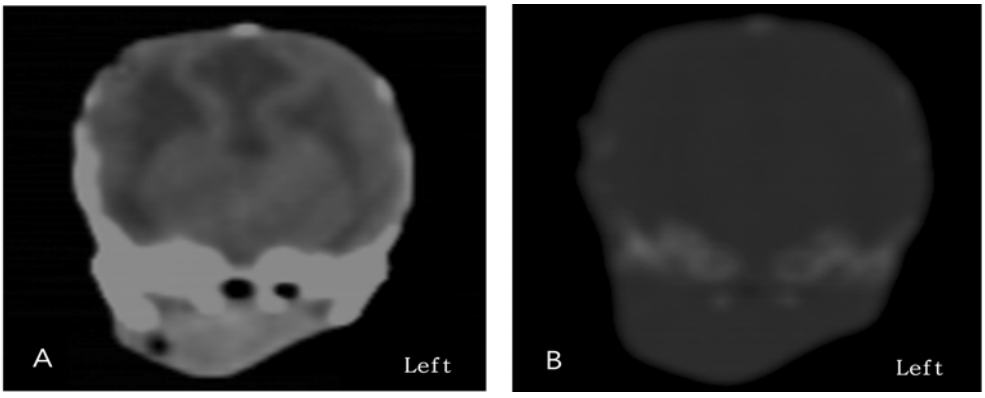


**Fig 3.** Transverse CT images of the skull. Asymmetrical dilatation of the lateral ventricles and ICAC appear as hypodense region on soft tissue window (A). The bone window shows absence of calvarium, frontal, parietal, and some occipital bone (B).

occipital bones(8). Sonographically, the acalvaria appears as an absence of cranial bones, with the presence of normal facial features(3). In addition it appears as an absence of shadowing posterior to the head structures, due to the absence of the bony cranium(3). Cerebral hemispheres should be visualized, though they may not appear in normal. The overlying skin should be seen surrounding the brain matter(3). A transverse CT scan of the brain can notice the absence of the calvarial bones with or without abnormal intracranial contents, such as ventricle enlargement, exencephaly, encephlocele, and so on.

Differential diagnosis for acalvaria include severe osteogenesis imperfecta and congenital hypophosphatasia(3). These pathologies provide inadequate visualization of the cranial bones due to the lack of ossification(3). In addition, these disease can be present with four limb abnormalities(7). Anencephaly is another differential diagnosis list, but the presence of cerebral tissue should negate this diagnosis(3). An encephalocele is possible, but the herniation of brain contents usually occurs through a smaller cranial defect(3). Exencephaly can be distinguished from acalvaria by the large amount of disorganized brain tissue protruding out of the base of the skull(3). Additionally hypocalvaria, the bones of the membranous neurocranium are present, but hypoplastic, abnormality is confusing with acalvaria(7,11). Fortunately Barr and Cohen devised "kidney-skull connection" during embryogenesis in an attempt to explain hypocalvaria in fetuses of women with angiotensin converting enzyme inhibitor use. Mostly the renal tubular agenesis was concurred with hypocalvaria(7,11).

The present report describes diagnosing acalvaria through physical examination, imaging modalities, and histopathological examination. The dog had shown absence of skull palpation, asymmetry of her face, and hypertelorism. Radiographs showed absence of calvarium and ventricle enlargement and its asymmetry was assessed by US. CT revealed the definitive range of calvarial bones defect and concurrent cerebral diseases including hydrocephalus and ICAC. Agenesis of the calvarial bone, the musculature, and the dura mater was observed on the gross necropsy examination. Osteogenesis imperfecta and hypophosphatasia were excluded by detecting no limb abnormalities on radiographs. On the US and CT, cerebral hemispheres abnormalities, such as absence, disorganization, or herniation were not shown, hence anencephaly, exencephaly, or encephalocele were ruled out, respectively. Also, acrania which has absence of cranium with abnormal development of cerebral hemispheres could have been differentiated with US and CT examinations. Hypoplastic membranous neurocranium or hypocalvaria were possible diagnoses, however, there were no abnormal kidney lesion on both gross and microscopic histological examination.

In human medicine, the reports of the acalvaria are rare, especially for primary acalvaria. To our knowledge, there has been no report about acalvaria in veterinary medicine field. This could be suspected by several reasons. It develops in rare incidence. Additionally, their abortion, stillbirth, or their early death can be contributed to their scarce detection.

Diagnosing acalvaria can be evaluated with imaging modalities successfully. Radiographs may fail to show the defects since the beam will not be in the place of the defect tangentially. Thus, there is a possibility that they could represent false negative findings. Also young dogs which have immature skeleton can be easily taken overexposure radiographs that cannot be imaged appropriately. CT, however, warrants precise and convenient assessment of location and shape of the defects. Anatomic localization of an abnormality is therefore more accurate in a tomographic image than in a conventional radiograph(1). CT study is useful for showing the extent of skull defect and its associated anomalies(9).

More reports and investigation would be required for the advanced research about skull defect in veterinary medicine. Furthermore relationship between skull defect and cerebral abnormalities, such as neural tube defects, based on embryological malformations should be investigated in veterinary medicine field. Also the correlation between open fontanelle and specific breed development should be studied in the near future.

## References


1. Amy ST. Principles of computed tomography and magnetic resonance imaging. *In*: Donald ET. Textbook of Veterinary Diagnostic Radiology. 5th ed. Saunders Elsevier. 2007; 50-77.
2. Bianca S, Ingegnosi C, *et al*. Prenatal and postnatal findings of acrania. Arch Gynecol Obstet 2005; 271: 256-58.
3. Cherie R. Primary Acalvaria. J Diag Med Sono 2006; 22: 407-10.
4. Cheryl PH, Jeannette JT, John CC. Am J Med Genet 1993; 46: 694-99.
5. Howard EE. Prenatal development. *In*: Howard EE. Miller's Anatomy of the Dog. 3rd ed. W.B. Saunders. 1993; 32-97.
6. Howard EE. The skeleton. *In*: Howard EE. Miller's Anatomy of the Dog. 3rd ed. W.B. Saunders. 1993; 122-218.
7. Kathleen M, Raj PK, *et al*. Acalvaria and hydrocephalus: A case report and discussion of the literature. J Ultrasound Med 1999; 18: 783-87.
8. Khadikar VV, Khadikar AV, Nimbaklar AA, Kinnare AS. Acalvaria. Indian Pediatr 2004; 41: 618-20.
9. Leboucq N, Martinez PM, *et al*. Aplasia cutis congenital of the scalp with large underlying skull defect: a case report. Neuroradiology 1994; 36: 480-2.
10. Lorenzo DB, Cynthia AM, *et al*. Neural-tube defects. N Engl J Med 1999; 341: 1509-19.
11. Mason B, Michael C. ACE inhibitor fetopathy and hypocalvaria: The kidney-skull connection. Teratology 1991; 44: 485-95.
12. Shushovan C, Shizuo O, *et al*. Congenital frontal bone defect with intact overlying scalp. Child's Nerv Syst 1993; 9: 485-87.
13. Suresh C, Meng KL, Victor YY. Fetal acalvaria with amniotic band syndrome. Arch Dis Child Fetal Neonatal Ed.

# 치와와견에서 발생한 원발성 Acalvaria 증례

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**요 약** : 약 3주령된 80 g의 암컷 치와와견이 두부 촉진 시 두개골이 확인되지 않으며, 어미젖을 먹을 시 이상을 보이고 성장 저하의 증상을 지닌 채 내원하였다. 신체 검사에서 입과 두부의 좌측 기울어짐과 좌측 안구 각막의 손상을 확인하였다. 방사선 검사에서 전두골과 두정골의 골음영이 관찰되지 않았으며, 초음파 검사에서 측뇌실의 확장을 관찰하였다. 컴퓨터단층촬영 검사를 통해 전두골과 두정골의 결손, 뇌실의 확장, 지주막 낭종을 확인하였다. 환자는 내원 익일에 폐사하였다. 부검에 의해 원발성 acalvaria로 확정 진단하였다.

**주요어** : acalvaria, 두개골 결손, CT, US, 개.