Congenital Hemicerebral Anomaly in a Stranded Pacific Harbor Seal (Phoca vitulina richardsi)

Authors: Christy A. McKnight, Taylor L. Reynolds, Martin Haulena, Alexander deLahunta, and Frances M. D. Gulland
Source: Journal of Wildlife Diseases, 41(3) : 654-658
Published By: Wildlife Disease Association
URL: https://doi.org/10.7589/0090-3558-41.3.654
ABSTRACT: A stranded 5-month-old female Pacific harbor seal (Phoca vitulina richardsi) was presented displaying tachypnea and diminished lung sounds. No neurological abnormalities were noted. The animal was treated for verminous pneumonia, but died 2 wk later. Gross necropsy examination revealed a severe obstructive verminous pneumonia associated with large numbers of Otostrongylus circumlitus. In addition, the majority of the right cerebral hemisphere was absent, with hypoplasia of the left cerebellar hemisphere, absence of the right pyramid, and malformation of the right occipital bone. Histopathologic findings included multifocal thrombosis and inflammation of pulmonary arteries, verminous pneumonia, and mild vacuolation of the subependymal white matter in the third ventricle representing swelling of myelin sheaths and edema. This is the first report of a hemicerebral anomaly in a marine mammal.

Key words: agenesis, brain, congenital defect, hydranencephaly, Pacific harbor seal, Phoca vitulina.
tifocal subacute to chronic thrombosis of small, medium, and large pulmonary arteries, intra-alveolar and intrabronchial nematodes with mild to moderate accompanying pyogranulomatous pneumonia, and moderate bronchial epithelial goblet cell hyperplasia.

The occipital bone on the right side of the skull was distorted and approximately half the size of the contralateral bone (Fig. 1). Upon intracranial examination, the right side of the cranial cavity contained approximately 10 ml of free, clear yellow fluid and there was absence of the majority of the right cerebrum (Fig. 2). Portions of the right olfactory peduncle, caudate nucleus, and hippocampus were still present. The left cerebellar hemisphere also appeared smaller than the right hemisphere. The left cerebrum and right cerebellar hemisphere appeared normal in size. There was mild enlargement of the left lateral ventricle. All cranial nerves appeared to be paired with no grossly detectable atrophy. Examination of the midbrain revealed the absence of the right crus cerebri. Examination of the medulla showed absence of the right pyramidal tract. The crus cerebri and pyramid normally contain projection fibers from the cerebral neocortex. The absence of the right neocortex in this seal explains the absence of the right crus cerebri and pyramid. Histopathologic examination of the brain revealed multifocal scalloped areas of mild vacuolation in the subependymal white matter of the third ventricle with mild astrogliosis, consistent with edema and swelling of myelin sheaths.

Infection with the metastrongyle nematode, *O. circumlitus*, is well recognized as a common pathogen in harbor seals (Geraci, 1978). Heavy infestations cause obstructive bronchitis and bronchiolitis (Lauckner, 1985), with the possibility of secondary infections, resulting in pneumonia or pulmonary abscesses (van der Kamp, 1987). The clinical signs of respiratory disease and death of this seal from verminous pneumonia is typical for juvenile seals in this area (Gulland et al., 1997). The apparent lack of clinical neurological abnormalities in this case, however, was surprising. The reorganizational
potential of the developing brain allows immature animals of many species to sustain damage to large areas of the brain and show remarkably little functional deficits (Chugani et al., 1996). A case of a 6-yr-old boy with unilateral cortical malformation had magnetic resonance imaging (MRI) findings that revealed that the unaffected hemisphere retained motor control of both hands (Staudt et al., 2001). It is likely that neuronal development during fetal growth in the seal allowed for similar cortical representation, resulting in apparently normal neurological signs. Domestic animals are much more dependent on their brain stem for survival than the cerebrum. The same may be true for the harbor seal, and this seal still had a normal left cerebrum.

Syndromes that can result in severe loss of cerebral tissue include hemicerebral anencephaly and hemihydranencephaly. Anencephaly is the term used for failure of cerebral development with concurrent skull malformation, and is a relatively common finding in human feti (Birnbacher et al., 2002). However, to the best of our knowledge there is no report of a nearly complete absence of a cerebrum (i.e., hemicerebral agenesis) in any mammalian species. There is a report of hemicerebellar agenesis in a 38-yr-old woman found incidentally on MRI, with no concurrent clinical signs of cerebellar abnormalities on neurological examination (Erdongan et al., 2002). This case was believed to be the result of intrauterine destruction of the cerebellum.

Another congenital abnormality, hemihydranencephaly—complete or partial destruction of one cerebral hemisphere with transformation into a membranous sac containing cerebrospinal fluid—has also
been described in humans (Greco et al., 2001). Its development has been ascribed to a number of causes, including infections, irradiations, fetal anoxia, medica-
tions, and twin–twin transfusion, leading to a vascular disruption (Greco et al., 2001). Reports of cerebral hemiatrophy caused by multiple developmental venous anomal-
ies also exist (Uchino et al., 2001). This marine mammal case represents he-
mihydranencephaly rather than anenceph-
aly because of the presence of the right olfactory peduncle, caudate nucleus, and hippocampus, and the fluid-filled right side of the cranial cavity. The majority of the
absent cerebral tissue receives its blood supply from the right side of the cere-
bral arterial circle. In utero compromise of this vasculature could explain the exten-
sive right-side hydranencephaly in this animal. With either congenital vascular mal-
formation or vascular thrombosis, blood supply to brain tissues would cease, lead-
ing to tissue necrosis and eventual hydran-
encephaly. As a result of the prenatal loss of this cerebral tissue, the absence of any neocortical projection fibers accounts for the absence of the right crus cerebri and pyramid. Normally the right cerebrum has abundant neurons that project to the left cerebellar hemi-
thesis and eventual hydranencephaly. The absence of this vasculature could explain the extensive right-side hydranencephaly in this animal. With either congenital vascular mal-
formation or vascular thrombosis, blood supply to brain tissues would cease, lead-
ing to tissue necrosis and eventual hydran-
encephaly. As a result of the prenatal loss of this cerebral tissue, the absence of any neocortical projection fibers accounts for the absence of the right crus cerebri and pyramid. Normally the right cerebrum has abundant neurons that project to the left cerebellar hy-
poplasia. Although the leptomeninges and dorsal cerebral cortical tissue were not seen grossly in the right skull, and are usu-
ally developed in cases of hydranencephal-
y (Nau et al., 1979), it is possible that they
could have been so thin that they were
easily destroyed during the necropsy.

In domestic animals, a similar hydran-
encephaly occurs bilaterally from in utero infection of the fetus with the Akabane, bluetongue, and Cache Valley viruses (Summers et al., 1995). In the case of this harbor seal, no associated infectious eti-
ology was identified.

LITERATURE CITED


Chugani, H. T., R.-A. Muller, and D. C. Chugan-


Erdogan, N., E. Kocakoc, D. Bekar, and Oz-

Fordyce, R. E., and A. G. Watson. 1998. Vertebral pathology in an early Oligocene whale (Getaceae, ?Mysticeti) from Wharekuri, North Ot-


Greco, F., M. Finochiaro, F. Pavone, R. R. Tri-
filetti, and E. Parano. 2001. Hemihydran-


Gulland, F. M. D., K. Burek, L. Lowenstine, L. 

tus) infestation of Northern elephant seals (Mir-


Leipold, H. W., and D. Troyer. 1995. Chromo-
sonal and genetic disorders. In Textbook of veter-


nisms of disease: A textbook of comparative gen-
eral pathology, 3rd ed., D. O. Slauson and B. J.
Cooper (eds.). Mosby, Incorporated, St. Louis, Missouri, pp. 300–305.


Received for publication 16 June 2004.