Toshiba Poster (Clinical or Tehcnical Research) Second Prize

Linear Echogenicity of Lenticulostriate Arteries in Neonates: A Significant Cranial Ultrasound Finding

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Summary

The lenticulostriate arteries are blood vessels that supply the region of the basal ganglia and internal capsule. They are normally anechoic structures on cranial sonography. In newborns, the echogenic appearance of lenticulostriate arteries has been reported in the literature to be associated with intrauterine infections, asphyxia and in association with chromosomal aneuploidy. However not much is known about the natural history of this finding on cranial sonography in newborns.

This retrospective study analyses the associated features of linear echogenicity of lenticulostriate arteries in term and preterm newborn and reviews the literature of the previously published case reports with this finding. The study raises the question that further detailed studies are needed to elucidate the natural history of this significant cranial ultrasound findings in newborns.

Objective

To determine the prevalence of echogenic lenticulostriate arteries on cranial sonography in newborns and to study the associated features of this finding.

Methods

Retrospective data analysis from September 1991 to June 1998. During this period the maternal and neonatal charts of all infants who had a diagnosis of echogenic lenticulostriate arteries on their cranial scans were analysed.

Setting

Intensive and special care nurseries, The Royal Women's Hospital, Melbourne.

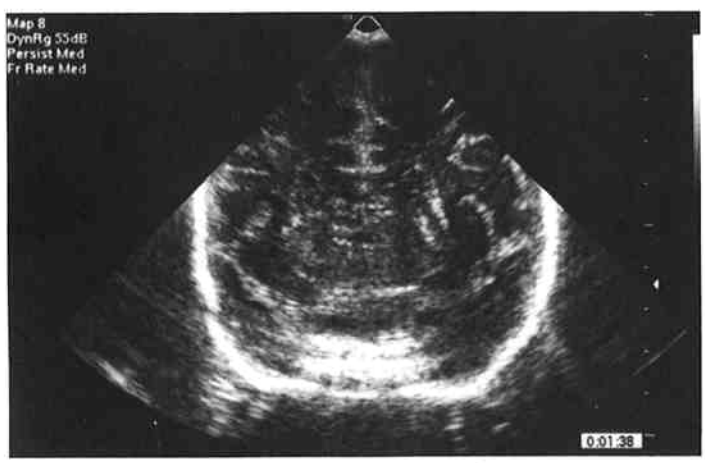

Figure 1 Coronal view of linear echogenicities seen in the region of the left basal ganglia and thalami.

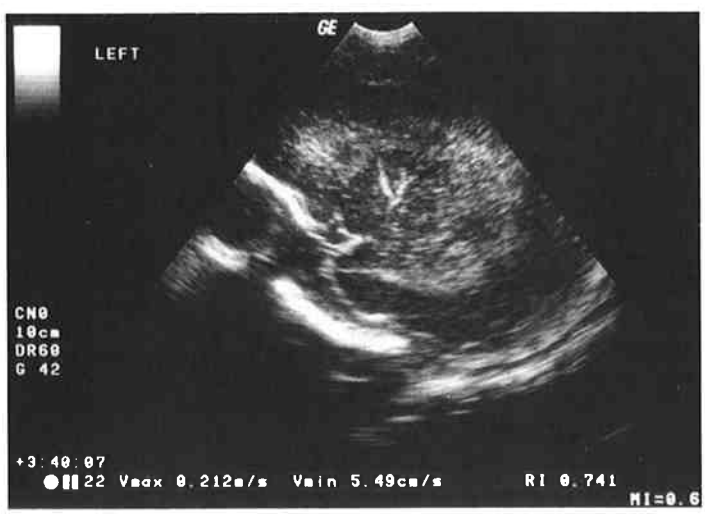

Figure 2 Parasagittal view of the branching linear echogenicities in the region of the basal ganglia and thalami corresponding to the lenticulostriate arteries.

In our unit routine cranial ultrasound scans are performed on all infants < 1500 grams birthweight and/or ≤32 weeks gestational age (GA). Apart from this scans are performed on newborns with craniospinal malformations, suspected or proven chromosomal abnormalities and infants with suspected meningitis, birth asphyxia and seizures. The finding of linear echogenic areas in the region of the thalami and basal ganglia (see figures 1 and 2) is further visualized by colour Doppler imaging. Once it is confirmed that the echogenic areas correspond to the region of the lenticulostriate arteries, a diagnosis of linear echogenicity of lenticulostriate (LS) arteries is made. This finding is always reported with a recommendation that the baby should have investigations to rule out viral infections. Maternal and baby serology for perinatal infections (cytomegalovirus, rubella, toxoplasmosis, herpes) and baby urine and nasopharyngeal aspirates for viral immunofluorescence and cultures are then performed and this constitutes "adequate" screening for perinatal infections.

Results

Ninety-two infants had evidence of linear echogenicity of lenticulostriate arteries. Eighty-three of these were bilateral

and 9 of them were unilateral. There were 40 females and 52 males. Table 1 describes characteristics of birth weight, gestation and age at diagnosis of these 92 infants.

Table 1 Characteristics of 92 Infants with echogenic lenticulostriate arteries on cranial ultrasound scan

	Median	Range
Gestational age (weeks)	30	23 - 41
Birthweight (grams)	1378	300 - 4290
Age at diagnosis (days)	24	0 - 118

Forty-seven out of these 92 infants were "adequately" investigated to rule out perinatal infections. Six out of 47 (13%) infants were confirmed to have positive infection: 5 of these infants were cytomegalovirus (CMV) positive; and 1 term infant had congenital toxoplasmosis (with hydrocephalus and periventricular calcifications along with echogenic LS arteries). However the 5 infants with CMV isolated from urine cultures had no obvious symptoms or any other clinical and ultrasound findings consistent with CMV disease.

There were 59 babies who were less than 33 weeks gestation at birth and therefore had cranial ultrasound in accordance with established unit protocol. Of these 27 babies were adequately screened for perinatal infections. Five of them were positive for CMV (18.5%).

Thirty-three infants out of 92 (38%) had the finding of echogenic LS arteries on their cranial scans at less than 7 days of life. In the remaining 59 infants, the diagnosis was made after 7 days of life.

In our study of 92 cases of linear echogenicities of the lenticulostriate arteries, the associated findings are variable. Only 6 of the 47 infants completely screened for perinatal infections had proven infection (5 CMV positive, 1 congenital toxoplasmosis). The different associations that were noted were Down syndrome (1), perinatal asphyxia (3), idiopathic neonatal seizures (3), craniospinal malformation (2), and culture proven bacterial sepsis (6).

Discussion

The first anatomical studies concerning the perforators of the carotid system were done by Duret[1] and Heubner[2]. The middle cerebral artery (MCA) gives origin to two main groups of deep perforators : **the lateral and medial lenticulostriate arteries**. Exceptionally the medial lenticulostriate artery may take origin from the anterior cerebral artery ACA. **The thalamotuberal artery** arises from the posterior communicating artery , but in a few cases may take origin from the MCA. The ACA gives rise to two groups of perforators: the **anterior lenticulostriate arteries and the recurrent artery of Heubner**. The internal carotid artery (ICA) gives origin to the **anterior choroidal artery** and to short direct perforators. Occasionally the anterior choroidal artery may take origin from the MCA. They supply the nuclei of the basal ganglia and the thalami and also the internal capsule.

Normally on cranial ultrasound in neonates, these vessels are inconspicuous within the grey matter nuclei and usually are indistinguishable from it. The first series of cases of echogenic vasculature in the basal ganglia was reported in 1988 by Teele et al[3]. In this report, the authors found only 12 cases over a 5 year period with echogenic "bright" vessels in the region of the basal ganglia and thalami and therefore reported this as an unusual finding. The source of this abnormality was believed to be vessels, specifically arteries, since the pattern was one of nonshadowing, branching, echogenic linear reflections.

Prior to this Grant et al[4], Rumack and Johnson[5], each described a patient with congenital CMV infection who had a prominent linear branching area of echogenicity in the basal ganglia and thalami and remarked that this finding suggested a vascular abnormality of unknown cause that would await neuropathological correlation.

Neuropathologic examination performed in 4 of 12 patients in the series of Teele et al[3] provided a histopathological correlate to the echogenic vessels and new evidence suggesting that mineralising vasculopathy , most likely following vasculitis, may be a feature of congenital CMV encephalitis. Five of these 12 cases had proven CMV infection, 2 rubella, 1 syphilis and 3 infants had Trisomy 13.

The authors raised the question then, "What is the cause of this mineralising vasculopathy?"[3]. Basophilic deposits (mineralisation) in arterial walls or hypercellular arterial walls are the likely reason for the echogenicity of these vessels on sonograms. Special staining with PAS, Prussian blue, von Kossa, and PTAH of specimens from one of their patients showed the deposits to be positive for iron, but negative for calcium. Thus one could postulate that babies who are exposed to an extra load of free iron could have an increased association with echogenic lenticulostriate arteries. In our study, 25 out of 92 infants (27%) were less than 1000 grams. These are infants who receive multiple transfusions in their neonatal period. Only 3 of these had a finding of linear echogenicity noted at less than 7 days. The remaining 22 had LS echogenicity noted after 7 days of life. Six of these infants had culture proven bacterial sepsis and 5 had CMV positive infection. Thus 11 of 92 infants (12%) were less than 1000 grams birth weight and had received multiple blood transfusions in their neonatal period. Apart from this feature there was no other specific association in these 11 infants to explain the echogenicity of lenticulostriate arteries on their cranial scans that appeared after 7 days of life.

The clinical information in this series supports the hypothesis that vasculitis is the instigating process. Necrotising vasculitis with subsequent mineralisation could be a non specific reaction to various insults. Spirochetal and viral infection of the central nervous system (CMV, rubella, HIV and other as yet undocumented viruses) may cause vasculitis in the fetus. Two of the 3 reported cases of Trisomy 13 in the series of Teele et al[3] had neuropathological finding of thick hypercellular arterial walls and mineralising vasculopathy in these arteries that were sonographically echogenic antemortem. The cause of sonographic and pathological vascular abnormality in neonates with this same chromosomal disorder remains unexplained.

Subsequently Ben-Ami et al[6] in 1990 confirmed by duplex sonography in 4 out of 11 patients the linear echogenicities in the basal ganglia and thalami in vivo to be arteries in the

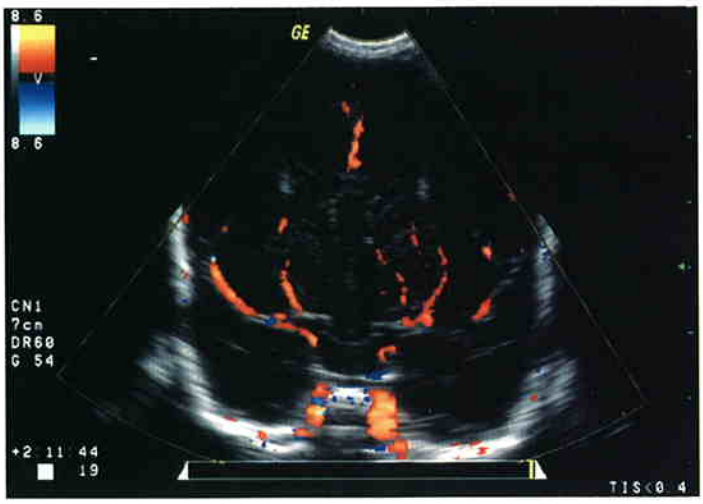

Figure 3 Colour Doppler imaging of the lenticulostriate arteies. (coronal view)

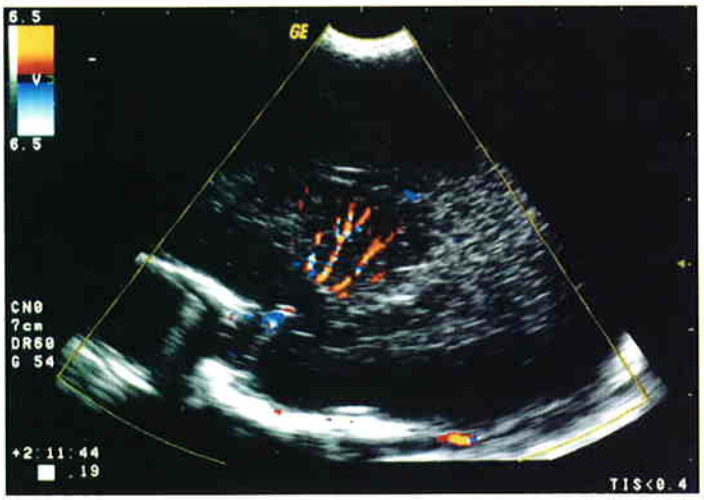

Figure 4 Colour Doppler imaging of the lenticulostriate arteries (sagittal view)

basal ganglia. They mentioned that the Doppler signal may be elicited in normal children and they are rendered vividly visible by colour Doppler (see figures 3 and 4).

Hughes et al[7] in 1991 presented a retrospective review of 25 patients out of 1324 patients who had areas of echogenicity in their thalami and basal ganglia that were of linear or branching linear distribution. Apart from cases of CMV and chromosomal abnormalities, they found 10 other patients in whom there were no clinical signs or suspicion of in utero infection; these patients had sustained a variety of anoxic, toxic and other injuries to the developing brain. Thus this series suggested a broader aetiological basis for linear areas of echogenicity in the basal ganglia and thalami.

In 1992, Weber K[8] et al described a series of 15 infants where hyperechoic lesions were seen in the basal ganglia and thalamic region were detected incidentally. Lesions showed a single punctate (n = 5), multiple punctate (n = 8), or stripe like pattern (n = 2). Thus linear stripe like echogenicity was seen in only 2 infants, one of which was detected on day 1 and persisted at one month of age; this infant had urine culture positive for CMV. The other case with birth asphyxia was noted to have this finding on day 1, but subsequently lost to follow-up. Their results indicated that hyperechoic lesions in the basal ganglia and thalamic region may be associated with congenital infections and asphyxia, but could indicate some other unknown pathology. No correlation was found between the morphology of foci and either clinical diagnosis or results of follow-up studies. They recommended Doppler sonography to evaluate possible vessel involvement.

Hyperechogenic areas in the thalamus and the basal ganglia (HTBG) were studied by colour Doppler flow imaging (CDI) in a series of 37 cases by Cabanas et al[9] in 1994. CDI confirmed that HTBG were allocated along the gangliothalamic vessels. Different patterns of HTBG observed were: punctate (n = 11), linear (n = 12) and mixed (n = 14). HTBG were associated with very heterogenous congenital disorders; however, a considerable proportion of patients also had uneventful neonatal periods. Although some infants presented with transitory disabilities (i.e: perinatal asphyxia, prematurity, respiratory distress syndrome), it was noteworthy that HTBG were always present soon after birth. Therefore, these data suggested that HTBG would most likely date back to fetal life.

Table 2 summarises the 5 published case series reports[3,6,7,8,9] of infants noted to have echogenic LS arteries and compares it to our data. Apart from the published case reports, the association of echogenic LS arteries and CMV infection in the fetus[10] and newborns with CMV and AIDS as isolated

Table2 Summary of case series reports of infants with echogenic areas in region of lenticulostriate arteries

First author	Year	No	Min age at D	Max age at D	MAJOR DIAGNOSIS CMV	Toxo	Rub	Aneuploidy	Asphyxia/ seizures	Others
Teele R[3]	1988	12	1	22	5	0	2	3	0	2
Ben-Ami T[6]	1990	11	0	6mo	2	0	2	1	0	6
Hughes P[7]	1991	25	1	3mo	4	0	0	3	5	13
Weber K[8]	1992	15	1	15mo	2	0	0	0	3	10
Cabanas F[9]	1994	37	0	150	1	2	1	1	3	29
Swaminathan M	1998	92	0	118	5	1	0	1	6	79

No	Number of cases with echogenic lenticulostriate arteries in each series
Min age at D	Minimum age at diagnosis
Max age at D	Maximum age at diagnosis
CMV	Cytomegalovirus
Toxo	Toxoplasmosis
Rub	Rubella

case reports[11,12,13] have also been described. These case series concluded that bright vessels in the basal ganglia and thalami on the sonograms of neonates should encourage the careful clinical and laboratory search for congenital infection or chromosomal abnormalities.

Conclusion

1 The exact pathogenesis of echogenic lenticulostriate arteries on ultrasound scan is still unclear and more systematic investigation is needed to unravel the natural history of this finding.
2 It is unclear whether presence of this finding on cranial sonography is suggestive of some antenatal event or is indicative of a post-natal evolving pathology.
3 The causative mechanism of deposition of iron in the vascular walls of these echogenic blood vessels needs to be further analysed.

Recommendations

The infants who demonstrate echogenic lenticulostriate arteries on cranial ultrasound scans need to be adequately investigated to rule out perinatal infections and until further evidence is obtained, biochemical studies to determine total body iron status especially in the infants less than 1000 grams birth weight need to be performed.

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ACUSON
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