

## CASE REPORT

# Portal vein thrombosis causing neonatal cerebral infarction

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Neonatal cerebral infarction often occurs in the absence of known risk factors. Two such cases are described in which portal vein thrombosis was documented during two dimensional echocardiography. In both cases, infarcts were consistent with embolic events. A novel mechanism is proposed, which may explain some cases of "idiopathic" neonatal cerebral infarction.

Neonatal cerebral infarction is a potentially serious, disabling condition. The incidence of such events has been reported to range from 1 in 4000<sup>1</sup> to 1 in 10 000.<sup>2</sup> These infants, usually asymptomatic at birth, present with seizures during the first few days of life. Current data suggest that 12–17.5% of neonatal seizures can be attributed to stroke,<sup>1,3</sup> making it the second most common cause of neonatal seizures.<sup>1,3a</sup>

The pathogenic mechanisms of neonatal cerebral infarction are complex. A number of predisposing factors have been implicated. Up to 25% of neonatal strokes have been reported in the literature as "idiopathic".<sup>4,5</sup> Some cases of "idiopathic" neonatal cerebral infarction have been attributed to a thromboembolic event, yet the source of the emboli remained unknown.<sup>3a,6</sup> We report on two full term neonates with embolic neonatal stroke and portal vein thrombosis. We propose a novel mechanism for neonatal cerebral infarction.

### CASE REPORTS

In the first case, a 3610 g boy was delivered at term to a 36 year old prima gravida mother. After a normal pregnancy, this was a spontaneous vaginal vertex delivery of a normal infant. Apgar scores were 8 at one minute and 9 at five minutes. At 24 hours of age, the infant had clonic movements of the left leg. Seizures continued over the next six hours, and the infant was transferred to our hospital. Neurological examination showed no abnormality other than mild left sided hypertonia.

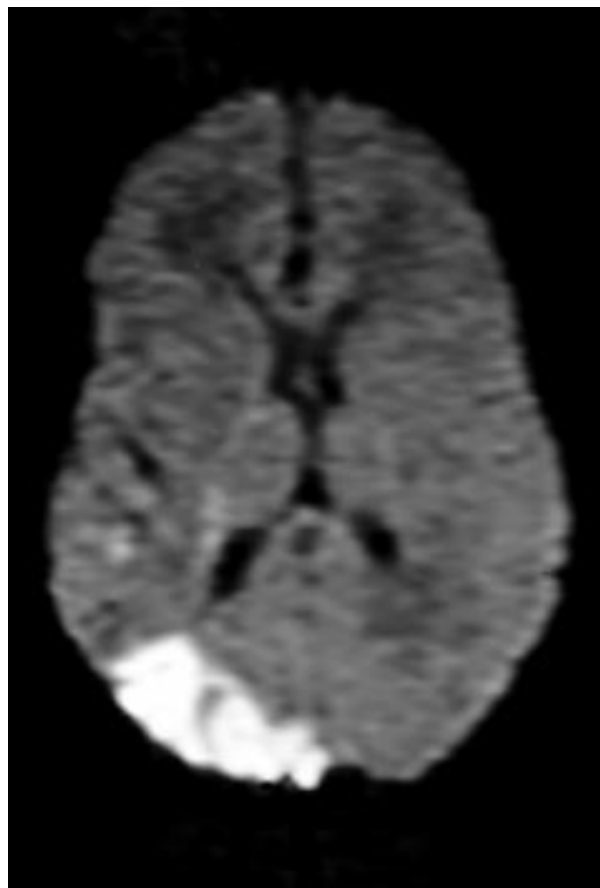
Magnetic resonance imaging identified acute infarcts in the right frontal and right parieto-occipital region (figs 1 and 2). The infarcts were consistent with an embolic event.

The second case was of a 4915 g girl delivered to a 29 year old prima gravida mother at 41 weeks gestation. After 37 hours of labour, low forceps and vacuum extraction were required during spontaneous vaginal vertex delivery. Bag and mask resuscitation were required for one minute. Apgar scores were 7 at one minute and 9 at five minutes. The infant was transferred to the well baby nursery and breast fed without difficulty.

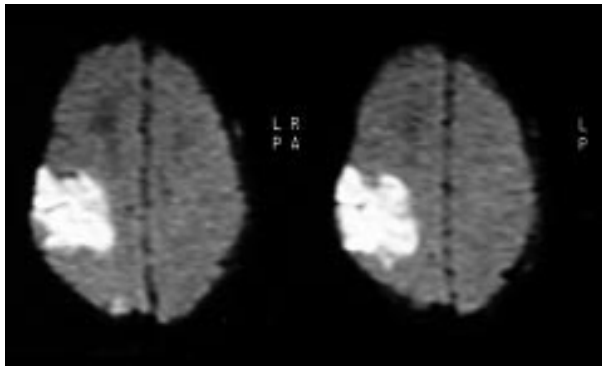
At 14 hours of age, a three minute episode of rhythmic twitching of the left hand and foot occurred. She was transferred to our hospital. Neurological examination was normal with no asymmetry of tone, power, reflexes, or brain-stem reflexes. A computed tomographic scan showed findings consistent with an embolic event (fig 3).

Laboratory studies in both cases were normal. Detailed coagulation studies included protein C, protein S, insulin receptor, partial thromboplastin time, activated protein C resistance, antithrombin III, and lupus anticoagulant. In case 1, cardiolipin antibody was normal; it was not measured in case 2. In both cases, echocardiography showed a small patent foramen ovale with left to right shunting. The heart valve leaflets were normal and there was no mural thrombus. In both cases, a portal vein thrombosis was noted during echocardiography. As part of our routine evaluation of neonates presenting with stroke, two dimensional echocardiography is performed to rule out right to left shunt lesions, mural thrombus, and valvular vegetative lesions.

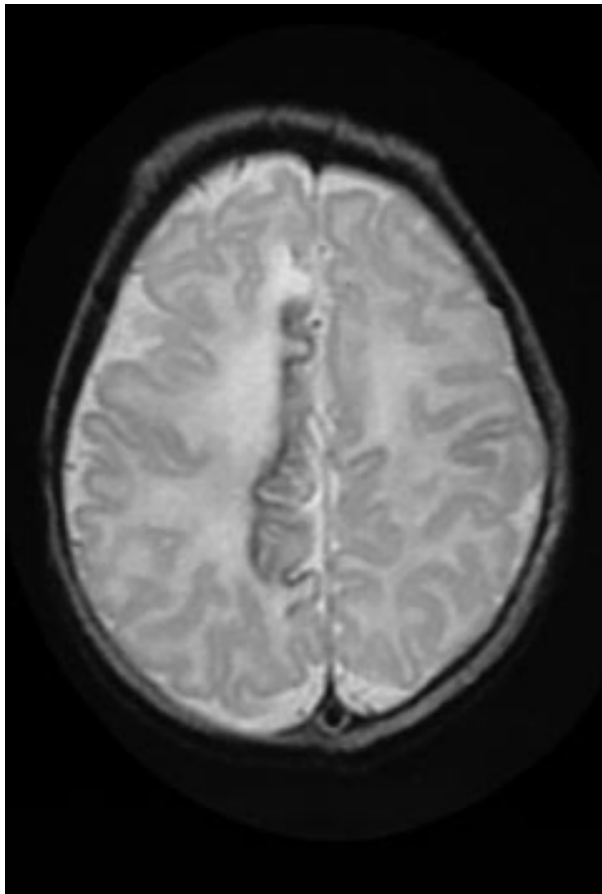
In our two cases, anticoagulation profiles were not undertaken in either mother. As both cases were referred from peripheral centres, data on placentae are not available.



**Figure 1** Magnetic resonance diffusion weighted image showing acute infarcts in the right parieto-occipital and right posterior frontal lobes involving cortex and subcortical white matter. T1 and T2 acquisition sequences showed very subtle abnormality only.



**Figure 2** Additional magnetic resonance diffusion weighted images from case 1 demonstrating acute cerebral infarcts.



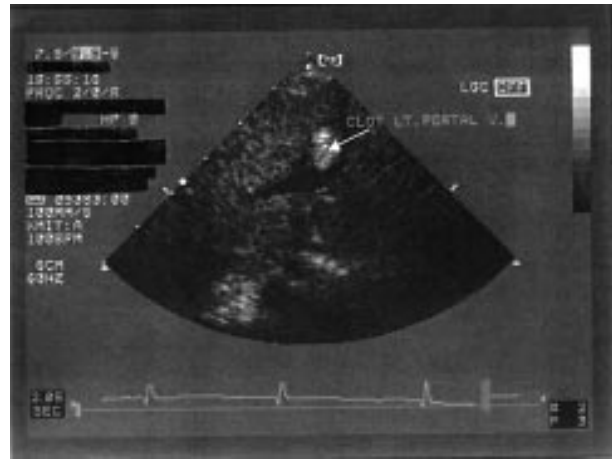
**Figure 3** Computed tomography showing right mesial frontoparietal high signal with low signal along cortex. Findings consistent with acute infarct of the right pericallosal artery.

As per our protocol, both neonates were appropriately anticoagulated using low molecular mass heparin (Enoxaparin) for three months. At follow up, both infants showed resolution of their portal vein thrombosis.

**DISCUSSION**

Risk factors for neonatal stroke have been identified in both retrospective and prospective studies. However, stroke among healthy full term neonates with no apparent risk factors has prompted much discussion and a search for possible mechanisms of pathogenesis.

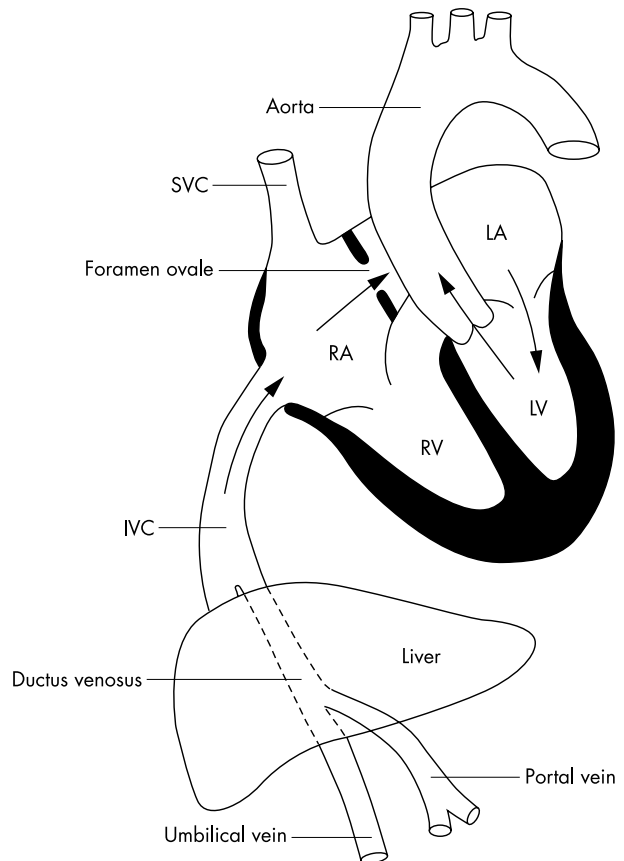
We have reviewed two such cases in which cerebral infarction of a thromboembolic nature occurred in two otherwise



**Figure 4** Two dimensional echocardiogram from the subcostal view showing thrombosis of the left portal vein.

healthy full term neonates. Neither neonate had any identifiable risk factors for portal vein thrombosis. The time course for onset of symptoms in both infants suggests that cerebral infarction was either a late intrauterine or perinatal event.<sup>1,6,7</sup> In both infants, portal vein thrombosis was detected on a two dimensional echocardiogram (fig 4), implicating this as the source of emboli.

The incidence of portal vein thrombosis in neonates is unknown. The most common cause is instrumentation, particularly umbilical vein catheterisation.<sup>8</sup> Other causes



**Figure 5** Normal fetal circulation. Paradoxical thromboemboli could travel from the portal vein to the systemic circulation as shown by arrows. LA, Left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava; IVC, inferior vena cava.

include sepsis, hyperviscous state, venous compression, and hypercoagulability.<sup>9,10</sup> In neither of our two cases were these risk factors present.

The unique characteristics of prenatal circulation provide a mechanism by which a thromboembolus could travel from the portal vein to the cerebral vessels of a fetus (fig 5). In the fetus, blood flows through the ductus venosus, which carries blood from the portal and umbilical veins to the inferior vena cava and right atrium. From the right atrium, most blood flows into the right ventricle and then to the main pulmonary artery. However, 27% of fetal cardiac output flows from the right atrium to the left atrium through the foramen ovale.<sup>11</sup> Therefore, emboli from a portal vein thrombus could pass through the ductus venosus and foramen ovale to enter systemic circulation. After birth, rapid closure of the ductus venosus eliminates this communication between the portal and systemic circulation.

Portal vein thrombosis with paradoxical emboli may help to explain the phenomenon of "idiopathic" cerebral infarction among healthy full term neonates. We suggest that abdominal ultrasound should be performed on neonates presenting with cerebral infarcts to rule out venous thrombosis as the source of thromboemboli.

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