

# Intrauterine fetal death of one of twins, coexisting with hydranencephaly in the surviving co-twin: A case report

\*J. A. Olowu<sup>1</sup>, I. A. Lagunju<sup>1</sup>, O. O. Tongo<sup>1</sup> and M. Atalabi<sup>2</sup>

Departments of Paediatrics<sup>1</sup> and Radiology<sup>2</sup>  
College of Medicine, University of Ibadan/ University College Hospital,  
Ibadan, Nigeria

E-mail: ilagunju@yahoo.co.uk, ilagunju@comui.edu.ng

## Summary

Hydranencephaly, a relatively rare malformation of the brain, is characterised by absence of the cerebral hemispheres and their replacement by sacs filled with cerebrospinal fluid.

It is one of the recognised forms of intracranial malformations associated with intrauterine fetal demise of one of twins in monochorionic twin gestation. This report illustrates the development of hydranencephaly in a surviving twin sequel to intrauterine fetal demise of the co-twin.

**Key words:** Hydranencephaly, Twins, Intrauterine fetal death.

## Résumé

Hydranencéphalie, une malformation du cerveau relativement rare, est caractérisée par l'absence d'hémisphères cérébraux et leur remplacement par des sacs remplis avec le fluide cérébrospinal. C'est l'une des formes connues des malformations intracrâniennes liées à la demise fœtale intrautérine de l'un des jumeaux dans la gestation monochorionique de jumeau. Cette communication illustre le développement d'hydranencéphalie chez un jumeau survivant à la suite de la demise fœtale intrautérine du co-jumeau.

## Introduction

Hydranencephaly is a congenital malformation of the central nervous system in which the cerebral hemispheres are missing.<sup>1</sup> It is associated with a total or near total destruction of the cerebral cortex and the basal ganglia, the thalami and lower brain centres are typically preserved.<sup>1,2</sup> It is a rare condition occurring in about 1 per 10,000 births or less, and found in 0.2% of infant autopsies<sup>3,4</sup>. Hydranencephaly is characterised by absence of the cerebral hemispheres and presence of a sac-like structure containing cerebrospinal fluid (CSF), surrounding the brain. Facial features are also uniformly normal and this distinguishes hydranencephaly from other major central nervous system anomalies.<sup>5</sup>

Hydranencephaly is considered to be the result of a destructive process or lesion in a previously normal brain.<sup>6</sup> Various theories have been proposed with regards to the aetiology of hydranencephaly. These include:

- Bilateral occlusion of the supraclinoid segment of the internal carotid arteries resulting in severe ischaemia,

infarction and damage to the intracranial structures supplied by these vessels.<sup>7,8</sup>

- Intrauterine infections by *Toxoplasma gondii*, *Cytomegalovirus*, and *Herpes simplex virus*, causing necrotizing vasculitis or encephalitis<sup>9,10</sup>
- Release of thromboplastic material from a deceased co-twin with resultant liquefaction of the normal brain tissue in the surviving twin.<sup>11,12</sup>

An increased incidence has been reported in smoking mothers.<sup>13</sup> An infant with hydranencephaly may appear normal at birth, the infant's head size and spontaneous reflexes such as sucking, swallowing, crying and limb movements may all appear normal.<sup>1</sup> However, as the child grows, the neurological deficits become evident. These include irritability, generalised spasticity, seizures, deafness, visual impairment, intellectual deficits and failure to acquire the expected developmental milestones.<sup>1</sup>

This report illustrates the development of hydranencephaly in a previously normal fetus, one of a set of twins, following intrauterine fetal death of the co-twin.

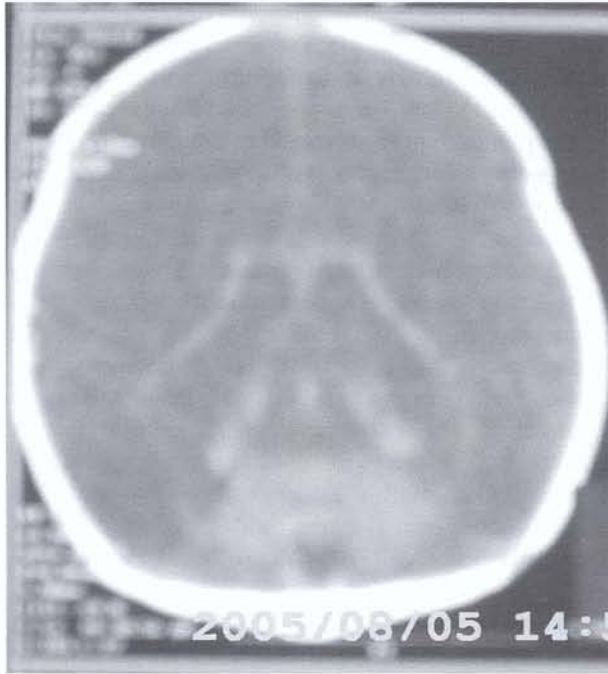
## Case report

Baby BT, a preterm female infant was delivered to a 33year old G2P<sup>1+0</sup> mother at a gestational age of 34 weeks. The mother was neither hypertensive nor diabetic; she was not a smoker and had never ingested alcohol. She booked for routine antenatal care at a tertiary health centre at a gestational age of 23 weeks. There was no history of exposure to irradiation in pregnancy. The only drugs taken in pregnancy were routine haematinics; oral Ferrous Gluconate 200mg thrice daily and oral Folic acid 5mg daily. The mother had no febrile illness or exanthematous skin rash in pregnancy.

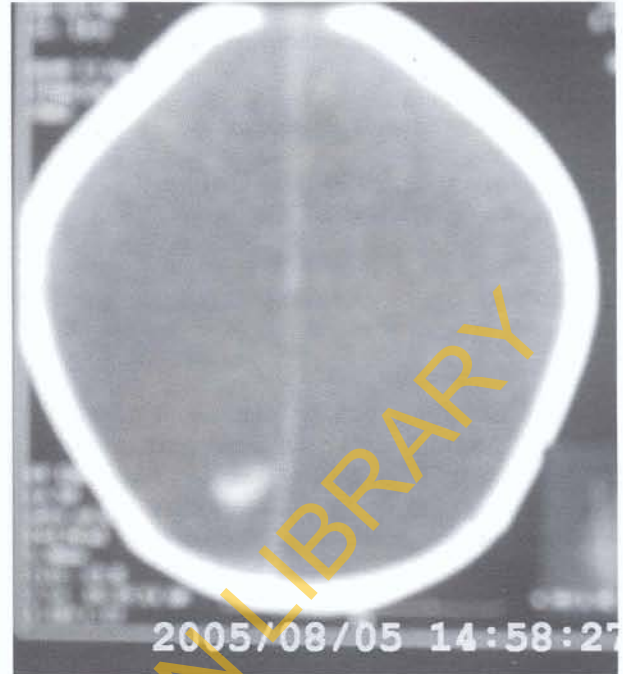
The first abdominal ultrasound in the index pregnancy was when she booked at 23weeks of gestation. This confirmed monochorionic twin gestation, however only one of the fetuses was alive while the other was found to have died. The living fetus was found to be normal with no abnormalities of the intracranial structures and no other congenital malformations. The pregnancy was therefore managed conservatively in order to allow for maturity of the surviving twin by the time of delivery. She continued on routine haematinics and twice weekly monitoring of her clotting profile. Fetal well being was monitored by fetal kick charts and was found to be normal.

A repeat abdominal ultrasound at the 31<sup>st</sup> week of pregnancy revealed abnormal intracranial structures in the

\*Correspondence



**Figure 1** Axial CT scan of the brain. Note the absence of the cerebral hemispheres and their replacement by fluid. There is also an absence of a circumferential rim of cerebral tissue which is usually seen in hydrocephalus. The posterior fossa contents are preserved.



**Figure 2** Axial CT scan of the brain showing absence of the cerebral hemispheres and an intact falx cerebri.

surviving twin. The ultrasonographer made an impression of ventriculomegaly. Baby BT's mother subsequently had the abdominal ultrasound repeated twice, at the 32<sup>nd</sup> and the 33<sup>rd</sup> weeks of pregnancy; these all confirmed the presence of abnormal intracranial structures. She was therefore delivered by elective Caesarean section at 34 weeks of gestation. BT, twin 1, had a birth weight of 2.15kg and Apgar scores of 7 and 10 at 1 and 5 minutes respectively. The second twin was a macerated still birth, which weighed 400grams.

On examination, BT was found to be a preterm female neonate, pink in room air, with an axillary temperature of 36.5°C. Her facial features were normal. Neurological examination revealed a conscious, active neonate, occipitofrontal circumference was 32.5cm, (within normal limits) and the anterior fontanelle was normotensive. The primitive reflexes: sucking and grasp reflexes were present and normal. Her pupils were equal and bilaterally reactive to light and muscle tone was normal in all limbs. Examination of the other systems revealed no abnormalities.

A transfontanelle ultrasound scan of the brain was carried out on the fourth day of life but the findings were inconclusive and a computerised tomography (CT) scan of the brain was thus suggested for further evaluation. This was done on the 10<sup>th</sup> day of life and findings were consistent with a diagnosis of hydranencephaly. Figure 1 is an axial CT scan of the brain showing a complete absence of the cerebral hemispheres, their replacement by fluid and preservation of the posterior fossa structures, all in keeping with a diagnosis of hydranencephaly. Figure 2

also demonstrates a complete absence of cerebral hemispheres and an intact falx cerebri. Baby BT's parents were counselled as to the prognosis of the condition and the minimal risk of recurrence in subsequent pregnancies. She was followed up at the high-risk neonatology clinic and was last seen at the age of 3 months. Evaluation at that time revealed generalised spasticity, signs of visual impairment and failure to attain any of the developmental milestones expected for her age. Her occipito-frontal circumference was however within normal limits for age.

#### Discussion

Hydranencephaly is a rare malformation of the brain. This condition is associated with a poor prognosis and most of the affected children do not survive beyond the first year of life.<sup>1,14</sup> A few cases of prolonged survival beyond the first year of life have been reported.<sup>14,15</sup> However, prolonged survival in these cases has not been associated with an improvement in responsiveness or awareness of self and environment.<sup>14,15</sup>

Loss of cerebral tissues supplied by the anterior circulation, that is, the internal carotid arteries, with preservation of the tissues supplied by the posterior circulation represents the hallmark of hydranencephaly. The thalami receive their blood supply from the posterior circulation perforating vessels, and are therefore preserved in cases of hydranencephaly.<sup>7,8</sup>

Thromboplastic material from a deceased co-twin has also been described as a cause of hydranencephaly in the surviving twin.<sup>11,12</sup> Hann et al<sup>11</sup> reported hydranencephaly

in a surviving twin following intrauterine fetal demise of the co-twin, while Larroche et al<sup>12</sup> reported classical cases of intracranial malformations in surviving twins coexisting with maceration of the co-twin in 7 sets of monozygotic twins. Although the pathogenesis of hydranencephaly in these cases is not fully understood, it is believed that transplacental transfer of thromboplastic material from the dead to the living fetus plays a major role.<sup>11,12</sup> In the report presented, an initial abdominal ultrasound scan confirmed twin gestation, with intrauterine fetal death of one of the twins at 23 weeks of gestation. Intracranial structures in the surviving twin were however normal at that time. A repeat abdominal ultrasound scan 8 weeks later revealed a major abnormality, initially considered to be hydrocephalus in the surviving twin. Further evaluation with a cranial CT scan however revealed a definitive diagnosis of hydranencephaly. This finding is consistent with previous observations which suggest liquefaction of previously normal brain tissue by thromboplastic material from a deceased co-twin as a potential cause of hydranencephaly. It is therefore probable that the intrauterine fetal death of the second twin resulted in hydranencephaly in Baby BT.

In the case presented, the availability of CT scan in this case facilitated the definitive diagnosis of hydranencephaly. The head size may be normal, increased or even smaller than the expected in infants with hydranencephaly. Cases of hydranencephaly with increased head size can be readily misdiagnosed as hydrocephalus.<sup>16</sup> However, the absence of a circumferential rim of cerebral tissue helps to differentiate hydranencephaly from hydrocephalus.

Intrauterine fetal death has been known to be associated with complications like sepsis, coagulopathy and secondary infertility in the mother. This report, like previous ones, suggests the development of hydranencephaly in the surviving fetus in cases of multiple gestation as another major complication.

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