A CASE REPORT OF ANTENATALLY DIAGNOSED CHIARI TYPE II MALFORMATION

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BACKGROUND

Chiari type II malformation is a common (1:1,000) congenital malformation of the spine and a small posterior fossa in form of myelomeningocele with associated descent of the brainstem and cerebellar tonsils, as well as other brain anomalies. As structural malformations are so variable, so are the long-term sequelae, such as cranial nerve palsies, neurogenic bladder, hydrocephalus and scoliosis.

This was an IVF donor egg and sperm pregnancy in a lesbian couple. It was an uneventful pregnancy up until 20 weeks gestation when at a routine antenatal scan a Chiari type II malformation with open myelomeningocele were noted. Baby boy was born in a good condition by an elective LSCS at 37+1 weeks gestation, weighing 2970 grams. The myelomeningocele was successfully repaired on day 1 of life. On post-operative imaging it was noted the baby had moderately enlarged lateral and third ventricles, with malformation of corpus callosum and absence of septum pellucidum. By 2 weeks of age a ventriculo-peritoneal shunt was inserted due to bulging fontanelle and irritability.

Information was collected from the patient, parents, notes and hospital databases.

OBSERVATION

The baby was referred to Community Paediatric clinic and was seen at 6 months of age for neurodevelopmental assessment and follow-up. He was found to be doing well and no longer requiring intermittent catheterisation. He was suffering with sleep apnoeas, snoring and excessive drooling, as well as cow's milk protein intolerance and reflux. Neurologically he was intact, with normal power, tone and reflexes of both upper and lower limbs and he was moving both of his arms and legs. However, he completely lacked head control. He had a good eye contact, normal hearing and a social smile.

KEY MESSAGE

The antenatal ultrasound scan remains the main means of diagnosis and initial assessment of this foetal malformation, which is followed by thorough postnatal evaluation with MRI scan.