

Images in Clinical Medicine

Dandy-Walker Malformation — A Rare Cause of Congenital Hydrocephalus

Received: April 3, 2018 Accepted: April 19, 2018

doi: <http://dx.doi.org/10.3329/jemc.v8i2.36737>

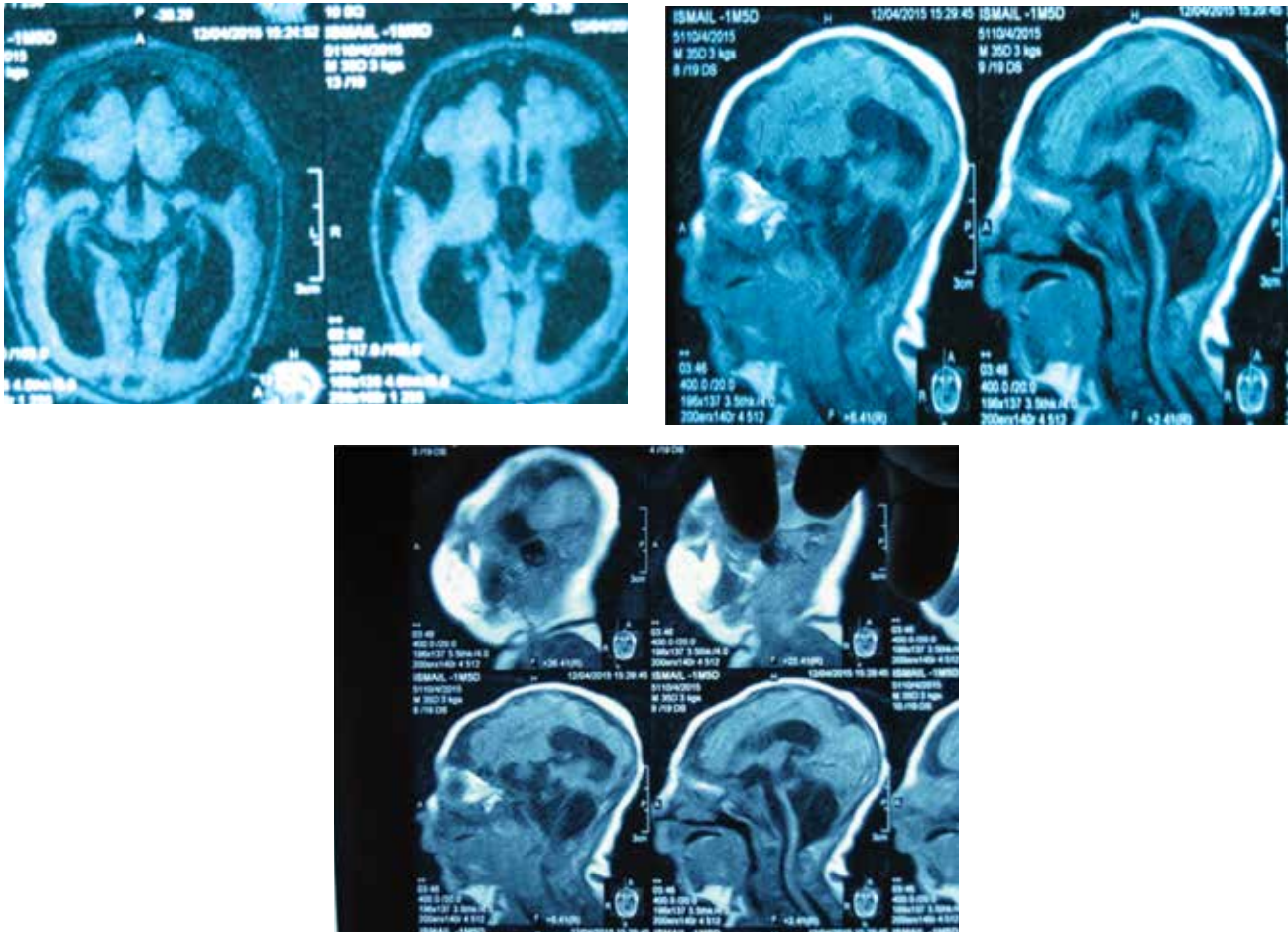


Fig 1. MRI of brain showing hypoplastic and everted cerebellum with a large posterior fossa cyst. Both lateral and 3rd ventricles are moderately dilated which is suggestive of Dandy-Walker malformation with moderate triventricular hydrocephalus with bilateral cerebral atrophy.

A one-month-old male infant was admitted in the Department of Pediatrics with the complaints of convulsion since birth. He was delivered at term by LUCS at hospital and had history of delayed cry after birth. Baby's mother was in irregular antenatal check-up and had no history of hypertension and diabetes. He is the 4th issue of nonconsanguineous parents. His two siblings died in infancy. His head circumference was 38.5 cm (50th percentile) while his weight and heights were normal. On examination baby was found less active, but vital signs were normal.

Neurological examination revealed spastic limbs. Other systemic examinations showed no abnormality. Investigations included a normal complete blood count and biochemistry profile. USG of brain showed that posterior fossa was enlarged with a large cystic lesion in the posterior fossa, which was a Dandy-Walker cyst. Dilatation of the ventricle was also evident. MRI of brain showed hypoplastic and everted cerebellum and a large posterior fossa cyst. Both lateral and 3rd ventricles were moderately dilated which is suggestive of Dandy-Walker malformation with moderate triventricular

hydrocephalus with bilateral cerebral atrophy. The baby was treated with anticonvulsant drugs and sent to neurosurgery department for further management.

Dandy-Walker malformation (DWM) is a rare abnormality of the central nervous system (CNS) and classically described as triad of hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle and enlargement of the posterior fossa. The incidence of DWM is one in 25,000 to 35,000 live births with a slight female predominance.¹ It accounts for 1–4% cases of antenatally detected hydrocephalus.² The Dandy-Walker complex is a continuum of aberrant development of the posterior fossa that has been associated with multiple congenital anomalies, radiographic abnormalities, and developmental delay.³

Although its pathogenesis is not completely understood, there are several genetic loci related to DWM as well as syndromic malformations and congenital infections.⁴ It may be difficult to identify DWM early due to lack of distinctive symptoms or signs. Progressive macrocephaly secondary to hydrocephalus (70–90%) may result in early recognition.⁵ There are several abnormalities associated with Dandy-Walker malformation like several extracranial anomalies, particularly facial and cardiac defects. The most common anomalies are ventriculomegaly (70%) and cardiac defects.⁵

In one retrospective series, cardiac anomalies were found in 42% of DWM patients. Reported congenital heart defects included ventricular septal defect, atrioventricular septal defect, atrial septal defect, pulmonary stenosis, and patent ductus arteriosus.² Midline defects such as agenesis of the corpus callosum, encephalocele, polycystic kidney disease and facial dysmorphism etc may also be associated with this condition. Infants with DWM may present with early signs such as vomiting, sleepiness, irritability, convulsions, unsteadiness and lack of muscle coordination.⁶

Hydrocephalus plays an important role in the development of symptoms and neurological outcome in patients with DWM. Imaging modalities, especially magnetic resonance imaging are crucial for the diagnosis of DWM and distinguishing this disorder from other cystic posterior fossa lesions. The aim of surgical treatment is to control hydrocephalus and the posterior fossa cyst.⁷

Overall mortality rate of DWM is 12–50%. Associated congenital abnormalities contribute to 83% of postnatal deaths and subnormal intelligence is reported in 40–70% of cases. The isolated Dandy variant abnormality has the highest incidence of survival.⁴

Prognosis for normal intellectual development varies depending on the severity of the syndrome and associated malformations. Difficulties in learning occur in 35–70% of children with DWS. Many children with DWS can be mainstreamed at school. Pediatricians, pediatric neurologists, pediatric neurosurgeons, geneticists, physical therapists, and educational specialists are to follow these children systematically and work to ensure that the child is given the best opportunities to reach his or her full potential.

Lazina Sharmin

Assistant Professor, Department of Pediatrics
Enam Medical College & Hospital, Savar, Dhaka
Email: drlazina22@gmail.com

References

1. Pascual-Castroviejo I, Velez A, Pascual-Pascual SI, Roche MC, Villarejo F. Dandy-Walker malformation: analysis of 38 cases. *Childs Nerv Syst* 1991; 7: 88–89.
2. Cornford E, Twining P. The Dandy-Walker syndrome: the value of antenatal diagnosis. *Clin Radiol* 1992; 45: 172–174.
3. Sasaki-Adams D, Elbabaa SK, Jewells V, Carter L, Campbell W, Ritter AM. The Dandy-Walker variant: a case series of 24 pediatric patients and evaluation of associated anomalies. Incidence of hydrocephalus and developmental outcomes. *J Neurosurg Pediatrics* 2008; 2: 194–199.
4. Kumar R, Jain MK, Chhabra DK. Dandy-Walker syndrome: different modalities of treatment and outcome in 42 cases. *Childs Nerv Syst* 2001; 17: 348–352.
5. Donkelaar HJT, Lammens M, Wesseling P, Thijssen HO, Renier WO. Development and developmental disorders of the human cerebellum. *J Neurol* 2003; 250: 1025–1036.
6. Simpkins CJ. Ventriculoperitoneal shunt infections in patients with hydrocephalus. *Pediatr Nurs* 2005; 31: 457–462.
7. Correa GG, Amaral LF, Vedolin LM. Neuroimaging of Dandy-Walker malformation: new concepts *Top Magn Reson Imaging* 2011; 22(6): 303–312.