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# Hydrocephalus Due to Dandy Walker Syndrome: A Case Report

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## INTRODUCTION

Hydrocephalus literally defined as "water in the head", is not a disease entity but a pathologic condition in which there is enlarged cerebral ventricles unrelated to cerebral atrophy or dysgenesis. It results from imbalance of production and absorption of cerebro-spinal fluid (CSF ) or obstruction to the passage of CSF between its origin and point of absorption[1]. There are various causes of hydrocephalus which could be congenital or acquired.

The Dandy-Walker syndrome may be defined as a developmental anomaly of the fourth ventricle and cerebellum in which the foramen of Magendie is atretic with or without associated atresia of one or both foramina of Luschka thus resulting in obstructive hydrocephalus[2] The incidence of the Dandy Walker syndrome in hydrocephalic patients is quite rare; most of the patients were under the age of two years at the time they presented with symptoms [3].

Below is presented a case of Dandy Walker syndrome who first presented with symptoms of hydrocephalus at the age of eleven years.

## CASE REPORT

C. O. is an 11-yr-old girl who was referred to the Neurology clinic of the University College Hospital Ibadan from a private clinic with complaints of dull persistent headache of about one year duration, occasional dizziness and visual blurring. There was neither associated fever nor past medical history of hospital admission.

Systemic examination was essentially normal. Cranial Computerized Tomography showed dilatation of all the ventricles especially the fourth ventricle. There was associated deficiency of the cerebellar penduncles and the vermis. A diagnosis of hydrocephalus secondary to Dandy Walker syndrome was made.

A ventriculo-peritoneal shunt was done with the cranial catheter tip being in the third ventricle. She was booked for posterior fossa exploration but her parents opted for treatment abroad.

## DISCUSSION

Hydrocephalus secondary to atresia of the outlets of the fourth ventricle (the foramina of Luschka and Magendi) was first described by Dandy and Blackfan in 1914 and later by Taggart and Walker in 1942[1]. Since this developmental anomaly occurs relatively early in fetal development there is usually associated dysplasia of the cerebellar vermis[4].

The aetiology of Dandy-Walker syndrome is not known and there is no evidence to suggest a genetic origin. The age of presentation ranges from first few weeks of life to adulthood, though most of the patients present in the first two years of life. The patients that do present late usually have one or both foramina of Luschka being patent[3] as may have been the case in this patient.

The symptoms are non specific. In patients under the age of two years, the most common presenting complaint is progressive enlargement of the head often with no neurological symptoms[5]. After the age of two years the head size and configuration are not as striking as in the infantile group and may be normal. Symptoms of increased intracranial pressure such as headache, vomiting, irritability, lethargy are more prominent in the older age group[4] as in the present patient.

Many of the patients presenting after 2 years have a history of febrile episodes or mild

trauma immediately preceding the acute onset of symptoms. In these patients mild meningitis, subarachroid haemorrhage or oedema of the membrane is believed to obstruct completely the already compromised flow of ventricular fluid from the fourth ventricle to the subarachnoid system [5]. The patient here had no prior history of head trauma or febrile illness.

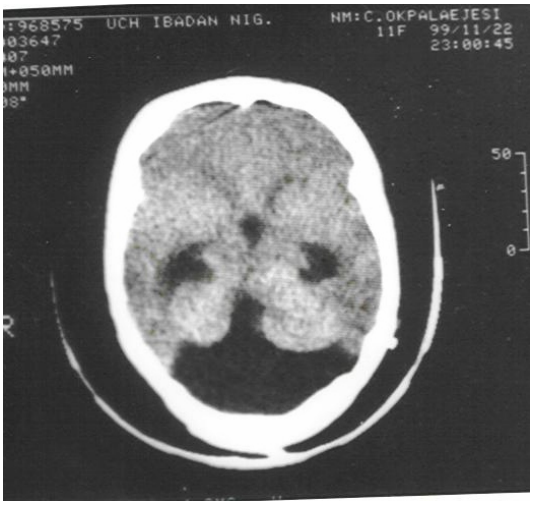
The patients below two years show cranio-facial disproportion in favour of the cranium. There is associated sutural diastases more marked in the lambdoid sutures. The so-called 'sunset' eye appearance may be seen. The posterior fontanelle is large and closes late. In the older child or adults the head may have a normal shape and size[3]

Ventriculography and/or angiography were done in the past to diagnose Dandy-Walker syndrome [2,4]. This has now been superseded by Computerized Tomography and Magnetic Resonance Imaging which show a large cystic mass occupying the posterior fossa. The pons and medulla are seen anteriorly with a thin rim of cerebellar tissue antero-laterally which is widely separated[6,7].

There may be associated congenital anomalies such as agenesis of the corpus callosum, encephalocele, porencephaly, cavum septum pellucidum and forking of the cerebral aqueduct[3]. The patient in this case did not have any of these associated anomalies.

The treatment consists of either cyst resection after posterior fossa exploration or creation of a ventriculo-peritoneal shunt with the cranial catheter tip in the third or lateral ventricle. An additional shunt tube may also be inserted into the fourth ventricle[4]. The patient here had a ventriculo-peritoneal shunt with tube tip in the third ventricle pending posterior fossa exploration.

***Fig. 1. A cranial computerized tomographic scan showing massive dilatation of the fourth ventricle. The third and the lateral ventricles are also dilated. The cerebellar hemispheres are also hypoplastic***

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