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Case Report

A CASE OF DANDY WALKER MALFORMATION WITH CONGENITAL DIAPHRAGMATIC HERNIA - A RARE VARIANT- CASE REPORT

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ABSTRACT
Dandy–Walker malformation is a rare abnormality of the central nervous system with a reported incidence of 1 in 25 000–35 000 live births and a slight female predominance. It accounts for 1–4% of cases of antenatally detected hydrocephalus. Dandy Walker variant forms part of the spectrum of Dandy Walker malformation. Congenital diaphragmatic hernia (CDH) is an uncommon; life threatening birth defect with incidence of 1:2000 to 1:3000 births.10% of cases may have CNS abnormalities. As per our knowledge, there is a single study about its association. Association between dandy walker malformation and congenital diaphragmatic hernia is very rare. We report a case where two associated congenital anamolies was found on autopsy in a stillborn fetus.

Key Words: *Dandy–Walker Malformation, Congenital Diaphragmatic Hernia, Stillborn*

INTRODUCTION
Dandy–Walker syndrome or Dandy–Walker complex is a congenital brain malformation involving the cerebellum and the fluid filled spaces around it. It is a disorder of ventral induction resulting in hind brain abnormalities incorporating varying degrees of hypoplasia of cerebellarvermis &/or enlargement of cisterna magna (Neil *et al.*,). Prevalence of the Dandy-Walker malformation among live births varies from about 1 in 25,000 to about 1 in 5000. About 45% cases are associated with chromosomal anomalies.

CASES
A female fetus stillborn at 26 weeks gestation to a gravida 2, para 1 mother, a product of second degree consanguineous parents was induced & submitted to autopsy.

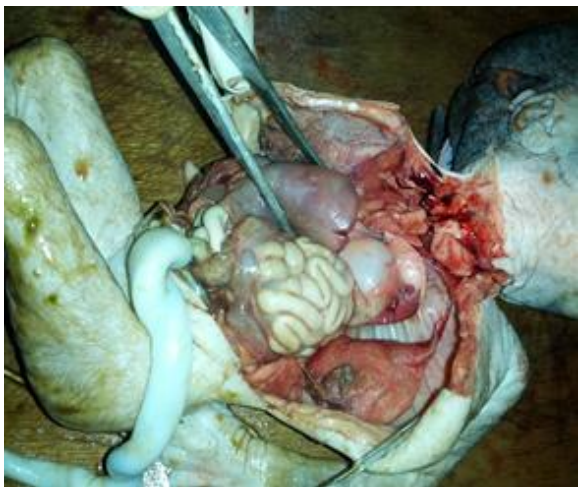

Figure 1: Showing left thorax (liver pulled towards right to expose stomach & spleen) with diaphragmatic herniation of liver, spleen & stomach

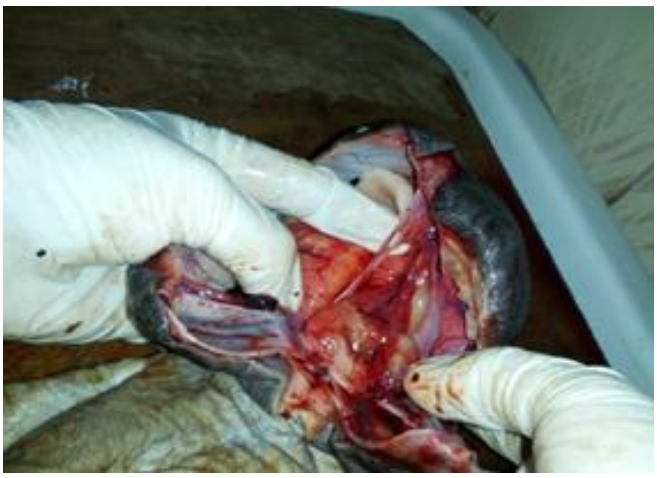

Figure 2: Showing posterior cranial fossa cyst with absent vermis

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On autopsy fetus had the following features

- Hydrocephalus
- Absent corpus callosum & vermis
- Cyst: adjacent to cerebellum in the region of fourth ventricle
- Diapghramatic herniation of liver, stomach & spleen into left hemithorax.
- Hypoplastic left lung.

DISCUSSION

Dandy–Walker syndrome or Dandy–Walker complex is a congenital brain malformation involving the cerebellum and the fluid filled spaces around it. The key features of this syndrome are an enlargement of the fourth ventricle; a partial or complete absence of the cerebellar vermis, the posterior midline area of cerebellar cortex responsible for coordination of the axial musculature; and cyst formation near the internal base of the skull. It is a genetically sporadic disorder that occurs one in every 30,000 live birth. The syndrome can appear dramatically or develop unnoticed (Deanna *et al.*, 2008). There is association between Dandy walker malformation and chromosomal abnormality in upto 45% cases. The severity of the diagnosis is based on the size of the posterior fossa, the presence of cystic dilatation of the fourth ventricle, and the degree of vermian hypoplasia. Simple enlargement of the posterior fossa with an enlarged foramen magnum and associated cisterna magna, which is known as "mega cisterna magna," is usually a benign finding when not associated with other congenital anomalies. At the other end of the spectrum are the well-known entity DWM, which is characterized by the presence of a large posterior fossa cyst with open communication between the fourth ventricles, absent or severely atretic inferior vermis, and enlargement of the posterior fossa with elevation of the confluence of sinuses, lateral sinuses, and tentorium. Extracranial malformations are also very common. Among those most frequently noted are facial hemangiomas, cardiovascular defects, and digital anomalies. The outcome for infants with the Dandy-Walker malformation varies widely, largely as a function of associated malformations and underlying diagnosis. Overall mortality for patients with the disorder is in the range of 27% with most deaths attributable to associated malformations, uncontrolled hydrocephalus, shunt malfunction or infection. Congenital diaphragmatic hernia (CDH) is an uncommon, life threatening birth defect with incidence of 1:2000 to1:3000 births, of which 96% are Bochdalek type. CNS abnormalities is seen in 10% cases. Association between dandy walker malformation and congenital diaphragmatic hernia is very rare.

Conclusion

Association between dandy walker malformation and congenital diaphragmatic hernia is very rare. We report a case where two associated congenital anamolies was found on autopsy in a stillborn fetus.

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