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Imaging in Posterior Fossa Malformations: A Pictorial Essay

KEYWORDS: Congenital heart disease, New born child, Radiograph Chest.

Reddy Ravikanth
MBBS, Post-graduate student in Radiology,
St. John's Medical College, Bangalore – 560034.

*** Partha Sarathi Sarkar**
MBBS, DMRD, DNB Senior Resident in Radiology,
St. John's Medical College, Bangalore – 560034.
* Corresponding Author

Ravi Hoisala
MBBS, DMRD, MD Professor in Radiology,
St. John's Medical College, Bangalore – 560034.

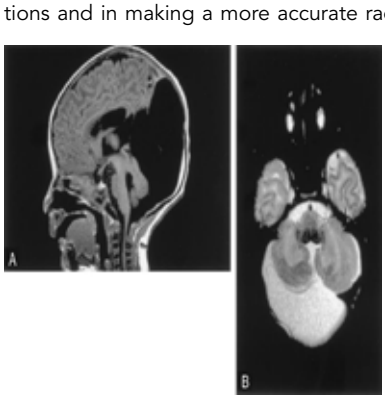
Babu Philip
MBBS, MD Professor in Radiology,
St. John's Medical College, Bangalore – 560034.

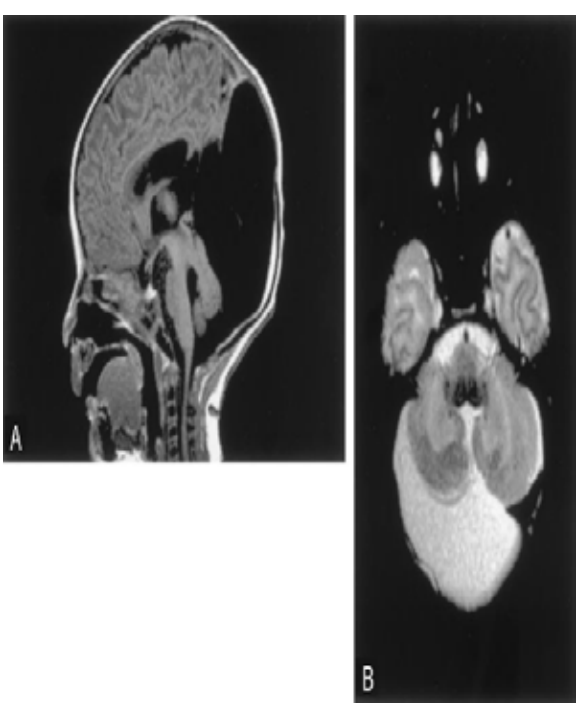
ABSTRACT *In twenty-first century medical science is advancing rapidly. It has helped to diagnose and treat nearly all the diseases making human to live a long and healthy life. Congenital heart disease is difficult to diagnose in womb*

due to many reasons like lack of expertise and lack of high end machines at all the diagnostic centers. Radiograph obtaining facility is available easily and hence can be used to diagnose congenital heart disease in a new born child thus helping quick intervention.

INTRODUCTION:

Cystic or cyst like malformations of the posterior fossa represent a spectrum of disorders, including the Dandy-Walker malformation, vermian-cerebellar hypoplasia, mega cisterna magna and arachnoid cyst. Differentiation of these lesions may be difficult with routine cross-sectional imaging; however, an accurate diagnosis is essential for proper treatment of planning and genetic counseling. Dandy-Walker malformation is easily diagnosed on the basis of the classical triad: complete or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and enlarged posterior fossa. Vermian-cerebellar hypoplasia is a general classification that describes congenital malformations with a normal sized posterior fossa, varying degrees of vermian and cerebellar hypoplasia and a prominent retrocerebellar cerebrospinal fluid space that communicates freely with a normal or dilated fourth ventricle. Mega cisterna magna can be asymmetric and can manifest apparent mass effect, simulating the appearance of an arachnoid cyst; therefore ventriculography or cisternography may be needed to demonstrate communication of the cystic mass with the subarachnoid space. A careful review of the embryologic development is essential in understanding these malformations and in making a more accurate radiologic diagnosis.





Dandy-Walker malformation with cerebellar dysplasia in an 8-day old neonate. Sagittal spin-echo image shows large posterior fossa CSF collection and dysplastic appearing cerebellar vermis. Axial spin-echo image shows abnormal folial pattern of cerebellar hemispheres.

Cerebellar hypoplasia in a 42-year-old male. Sagittal spin-echo image shows a small cerebellum

in a fluid-filled posterior fossa. The pons and medulla are abnormally small.

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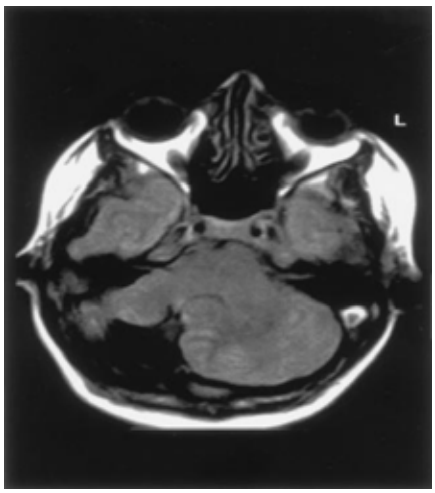
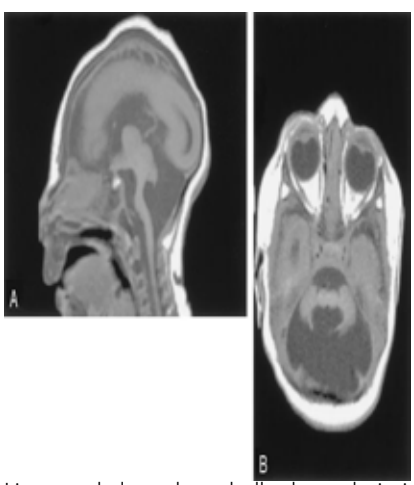
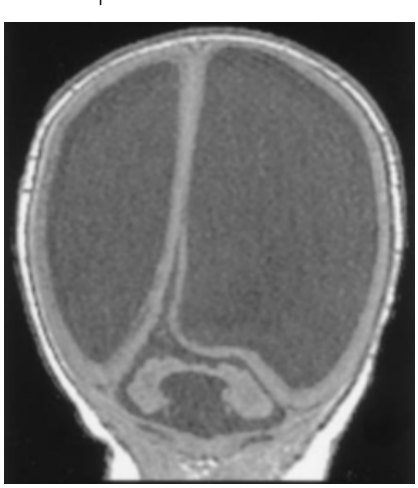


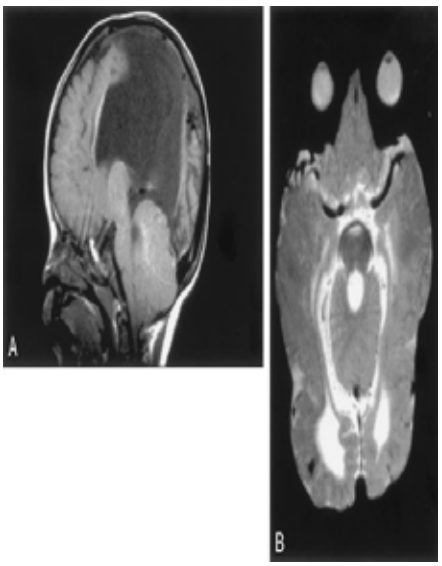
Unilateral cerebellar hypoplasia in 33-year-old female. Axial spin-echo image shows a small right cerebellar hemisphere. The left hemisphere and vermis appear normal.



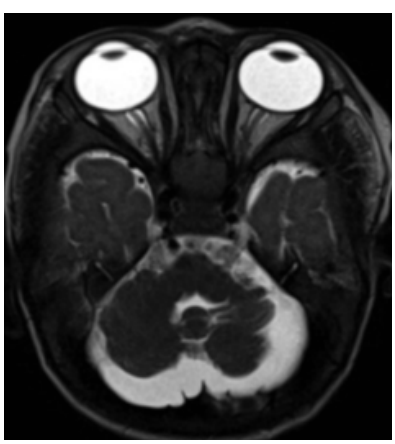
Lissencephaly and cerebellar hypoplasia in a 6-day-old neonate. Sagittal spin-echo image shows complete cerebral lissencephaly and a very small cerebellum. Axial spin-echo image shows the very small cerebellar hemispheres and the small pons.



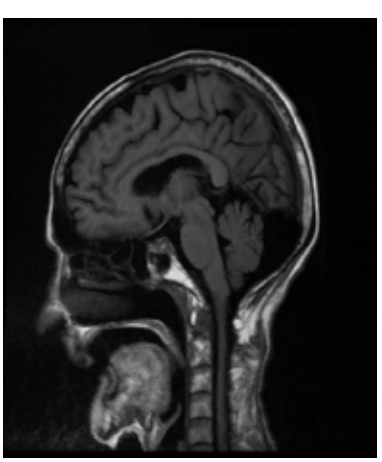
Coronal T1W spin-echo image in a neonate with diffuse cerebellar dysplasia showing marked ventriculomegaly and small posterior fossa.



Rhombencephalosynapsis in a 7-year-old male child. Sagittal spin-echo image shows abnormal cerebellar vermis. Axial spin-echo image shows continuity of the cerebral hemispheres across the midline without a midline cerebellar vermis.



Global cerebellar hypoplasia in a 2 year old child with congenital CMV infection.



Sagittal MRI image of a 33 year old female patient with giant cisterna magna with enlargement of the subarachnoid space in the inferior and posterior portions of the posterior

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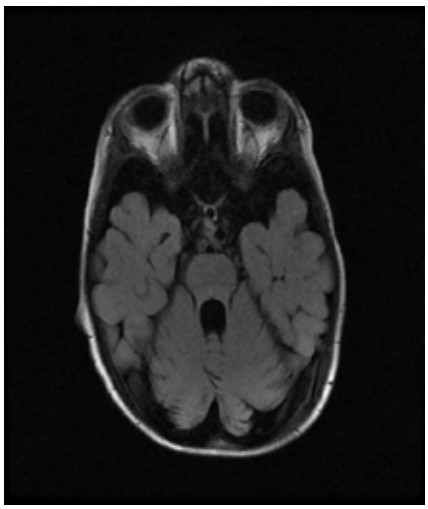
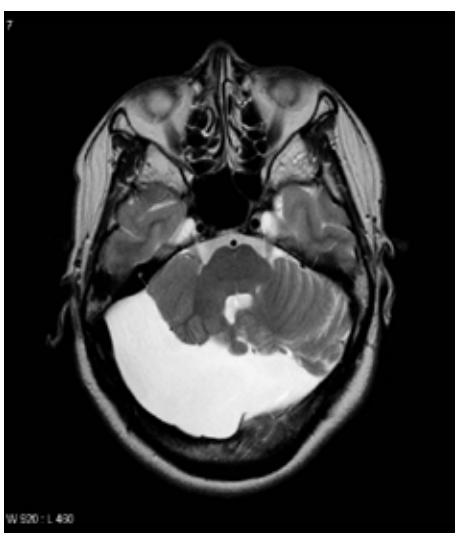
fossa measuring > 10 mm.



Axial MRI image of a 22 year old male showing the molar tooth configuration of mid brain – suggesting Joubert's syndrome.



Axial T2W spin-echo MR image of a 22 year old male demonstrating a posterior fossa arachnoid cyst. Arachnoid cyst shows CSF intensity.

CONCLUSION:

Cystic or cyst like malformations of the posterior fossa are often difficult to differentiate on the basis of their radiologic appearances alone. This spectrum of disorders includes Dandy-Walker malformation, vermian-cerebellar hypoplasia, mega cisterna magna, rhombencephalosynapsis and the development anomaly, arachnoid cyst. It is very important for prenatal counseling to recognize those syndromes associated with vermian-cerebellar hypoplasia that form confined nosologic entities and that are associated with a poor prognosis and high prevalence of genetic recurrence (eg. Joubert syndrome). The differentiation between mega cisterna magna and posterior fossa arachnoid cyst can be difficult on the basis of CT or MR imaging findings alone. Mega cisterna magna can be asymmetric and manifest with mass effect, imitating the appearance of an arachnoid cyst. Cisternography or ventriculography may be required to demonstrate or exclude communication of the cyst with the subarachnoid space, which is important in making the clinical decision of shunt placement.

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Informed consent: Obtained

Ethical clearance: Taken from Institutional Ethical Review Board, St. John's Medical College, Bangalore –