



## A Rare Coexistence of Partial Agenesis of Corpus Callosum and Diastematomyelia

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### ABSTRACT

*Corpus callosal agenesis (partial or complete) is a congenital malformation of the brain that is relatively rare and often associated with other anomalies of the central nervous system like Chiari malformation, Dandy Walker syndrome, lissencephaly and holoprosencephaly. Its association with a congenital spinal dysraphism called diastematomyelia is relatively rare. A case of coexisting partial agenesis of corpus callosum and diastematomyelia of the lumbar spinal cord, is reported here, in a 23 day old female neonate presenting with repeated attacks of seizures since birth.*

**Keywords:** colpocephaly, conusmedullaris, magnetic resonance imaging, spinal dysraphism

### INTRODUCTION

Corpus callosum is the major white matter commissure connecting the cerebral hemispheres. Partial or complete agenesis of corpus callosum is uncommon, occurring in 1 in 20000, and represents the consequence of an in-utero insult <sup>(1)</sup>. It is more often seen in males than females and appears to have an association with maternal alcohol consumption <sup>(1)</sup>. Diastematomyelia is another rare developmental anomaly of dorsal induction that occurs at around 4-5 weeks of intrauterine life,

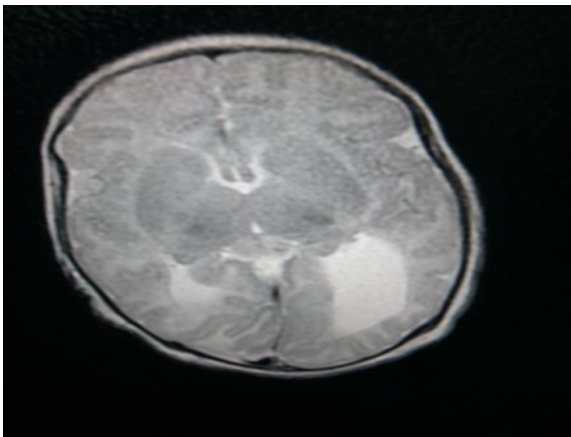
where the spinal cord is sagittally split into two hemicords, with (more common) or without an intervening fibrous or osseous septum <sup>(2,3)</sup>. A rare association of partial agenesis of corpus callosum with spinal diastematomyelia is illustrated here, in a neonate.

### CASE REPORT

A female neonate, 23 days old, was brought to the Department of Paediatrics with complaint of several episodes of generalised seizures since birth.

Maternal history was not suggestive of any prenatal insult and antenatal ultrasound conducted elsewhere was normal. Her physical examination was not striking. She was referred to the Department of Radiodiagnosis to undergo Magnetic Resonance Imaging (MRI) of the Brain.

Axial MRI scan revealed dilatation of occipital horns of lateral ventricles (colpocephaly) as well as abnormal T2 hyperintensity over bilateral periventricular white matter suggestive of periventricular leukomalacia (Fig.1).



**Fig 1:** Axial T2 weighted MRI Brain showing dilated occipital horn of lateral ventricles

Sagittal section scan further suggested absence of splenium and posterior aspect of the body of corpus callosum (Fig 2).



**Fig 2:** Sagittal T1 Weighted MRI Brain showing absence of posterior aspect of body and splenium of corpus callosum

She was thereafter advised MRI of the spine, to rule out other associated congenital anomalies. Examination of sagittal images of the lumbosacral region showed widening of the conus medullaris. Segmentation and fusion anomalies of the vertebral bodies were seen along T11 to L1 levels (Fig3).



**Fig 3:** Sagittal MRI Spine showing expansion of conus medullaris along with vertebral body fusion.

Axial scan demonstrated a longitudinal split of the lumbar spinal cord with the two hemicords separated by intervening cerebrospinal fluid (CSF) without evidence of any midline bony spur. (Fig 4)



**Fig 4:** Axial T2 Weighted MRI of lumbar spine showing the two hemicords with intervening CSF

She was prescribed antiseizure medications and advised regular follow up.

## DISCUSSION

The formation of the corpus callosum starts with the development of the genu during the 11<sup>th</sup> week of intrauterine life, the body, isthmus and splenium develop at a later stage and finally the rostrum. If

this normal developmental process is disturbed, the corpus callosum may be partially or completely absent. In partial agenesis, posterior aspect of the body and the splenium may be missing which can lead to isolated dilatation of posterior horns of lateral ventricles termed colpocephaly (due to lack of support from the splenium) and widely spaced lateral ventricles. These typical findings were observed in our case. A dilated high riding 3<sup>rd</sup> ventricle and a dorsal cyst may be evident (a feature not found in this patient). Isolated agenesis of the corpus callosum is usually asymptomatic but may present with seizures, mental retardation or cerebral palsy<sup>(4)</sup>.

Diastematomyelia, the second coexisting anomaly observed here, can be classified into two types<sup>(5)</sup>. Type I is a severe form presenting with neurological deficits in early adulthood, in which there is an osseous septum dividing the thecal sac into two; and Type II is of a milder variety, usually asymptomatic, where a solitary dural sac is present with or without the presence of a fibrous septum. Diastematomyelia Type I is more commonly associated with segmentation and fusion anomalies of the vertebral column as well as spina bifida, butterfly and hemi-vertebra<sup>(6)</sup>. Coincidentally, our case exhibited features of Type II (the absence of intervening osseous septum) but revealed fusion of vertebral bodies. The most common location of this spinal dysraphism is the thoracolumbar region followed by cervical spine.

Corpus callosum agenesis can be associated with other conditions, most commonly Dandy Walker syndrome, Chiari I and II malformation, Schizencephaly, Lissencephaly and holoprosencephaly. But an association with Diastematomyelia, as seen in our case, is rare. However, a study conducted by Erol F.S. et al revealed the presence of diastematomyelia in 16 out of 85 patients diagnosed with partial corpus callosal agenesis<sup>(7)</sup>. Hence it is postulated that MRI screening of the spine needs to be carried out in patients with corpus callosal agenesis.

## CONCLUSION

A diagnosis of Corpus callosal agenesis should be followed by investigation of the spine to rule out its probable association with spinal dysraphisms like Diastematomyelia, for optimum management of its neurological sequelae.

## REFERENCES

1. Kumar P, Burton BK. Congenital malformations, evidence-based evaluation and management. McGraw-Hill Professional. 1<sup>st</sup>Edn.(2008).
2. Van der Knaap M. S, Valk J. *Classification of Congenital Abnormalities of the CNS*. American Jour of Neuro Radiology 1988; 9:315-326.
3. Castillo M, Mukherji SK. Imaging of paediatric head neck and spine. Philadelphia: Lippincott Raven; 1996. pp. 630–35.
4. Christopher Verity, Helen Firth, Charles French-Constant. *Congenital abnormalities of the central nervous system*. J Neurol Neurosurg Psychiatry 2003;74(Suppl D):i3–i8
5. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation. Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery.1992;31:451–80.
6. Allen LM, Silverman RK. *Prenatal ultrasound evaluation of fetal diastematomyelia: two cases of Type I split cord malformation*. Ultrasound Obstet Gynecol. 2000;15:78–82.
7. Erol FS, Ozturk S, Akgun B, Cakin H, Kaplan M. *How innocent is corpus callosum dysgenesis?* Pediatr Neurosurg. 2013; 49(1):24-8.