Colpocephaly

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Colpocephaly was first described by Benda in 1940 and termed vesiculocephaly but was renamed colpocephaly by Yakovlev and Wadsworth in 1946.

Colpocephaly is a congenital enlargement of the occipital horns of the lateral ventricles. This is caused by a global impairment in brain development with aberrant migration of neuroblasts resulting in a decreased thickness of cerebral white matter in the posterior part of the sentrum semiovale. Hence the secondary dilatation of the occipital horns. This abnormality is thought to occur at 1 - 4 months' gestation.[1]

Associated CNS abnormalities include corpus callosum agenesis, neuronal migration disorders (lissencephaly, pachygyria), schizencephaly, microcephaly, meningomyelocoele and hydrocephalus. In corpus callosum agenesis often associated with Chiari II malformation the ventricles are also widely spaced, parallel and pointed anteriorly.

The clinical features may include learning disability, seizures, spasticity, hypotonia and visual abnormalities.

It has been postulated that certain aetiologies may predispose to colpocephaly. These are intra-uterine infection, IUGR, perinatal anoxic ischaemic encephalopathy, maternal drug ingestion and trisomy 8 and 9.[2]

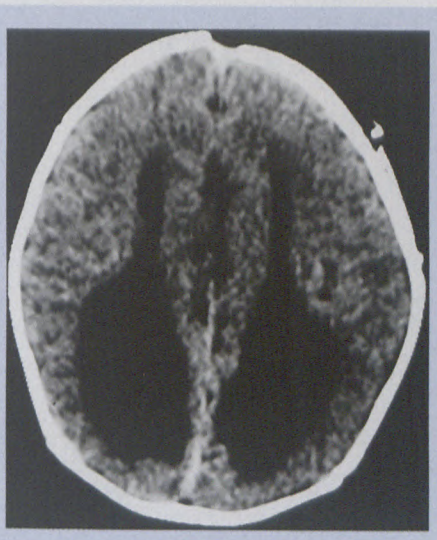

Fig. 1. Axial non-contrast CT scan of a child's brain with corpus callosum agenesis shows the dilated occipital horns of colpocephaly. The ventricles are also widely spaced and pointed anteriorly.

The diagnosis of colpocephaly is made on CT, MRI or ultrasound (Fig. 1).

References

1. Cerullo A, Marini C, Ceroli S, Carelli V, Montagna P, Tinuper P. Colpocephaly in two siblings: Further evidence of a genetic transmission. *Dev Med Child Neurol* 2000; **42:** 280-282.
2. Noorani P, Bodensteiner J, Barnes P. Colpocephaly: Frequency and associated findings. *J Child Neurol* 1988; **3:** 100-104.