

# Bilateral Perisylvian Syndrome- A Case Report

S TANEJA, AK CHATURVEDI

Ind J Radiol Imag 2006 16:2:169-171

**Key words :** - Bilateral Symmetrical Polymicrogyria, MRI

## INTRODUCTION

The term Congenital Bilateral Perisylvian Syndrome describes a structural malformation of the brain in which the underlying anomaly is Polymicrogyria, a malformation of the outer layer of the cerebral cortex. Polymicrogyria may have a focal or regional distribution or involve the whole cortical mantle. There are consequently wide spectrums of clinical manifestations, which include children with severe encephalopathies (brain impairments) and intractable epilepsy or normal individuals with selective impairment of cognitive functions (mental processes) in whom the mild cortical abnormality is only detected on pathological brain study.

We report a case of Bilateral Perisylvian Syndrome.

## CASE REPORT

A 15-Year-old girl presented with generalized tonic clonic seizures off and on since 1994. Born of a normal delivery in 1989 she had bilateral club feet with spastic paraplegia at birth. The child showed delayed milestones, slow cerebration and mental retardation. She was unable to walk and had atrophy below knees. There was dysarthria and drooling of saliva. On examination there were exaggerated deep tendon reflexes with extensor plantars. The patient was on anti epileptics since 1994.

A CT scan of the patient was done two years back, which showed deep B/L symmetrical clefts in the place of sylvian fissures

MRI of brain revealed multiple small gyri in bilateral parieto-occipital cortices. There was apparent thickening of the cortical white matter (Fig 1). Bilateral sylvian fissures were hypo plastic and were seen to extend dorsally up to the perirolandic region (Fig 2). B/L deep symmetrical clefts were seen in the place of sylvian fissures (Fig 3). Bodies of B/L lateral ventricles showed inverted appearance (Fig4).

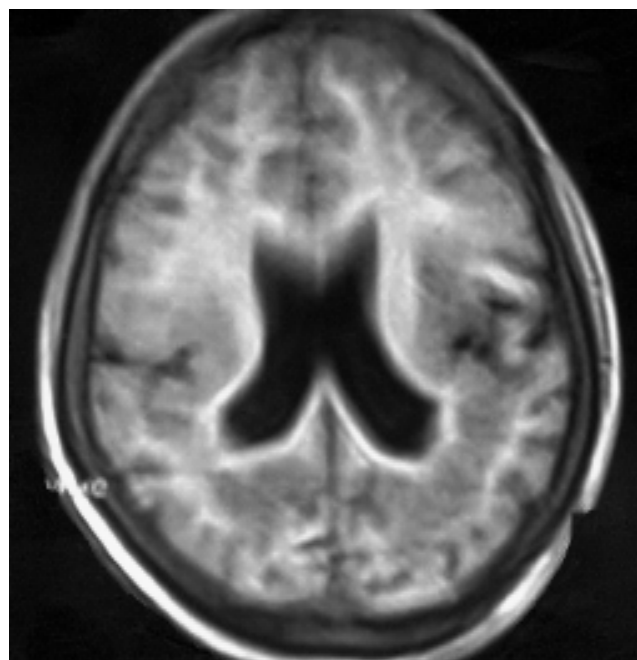


Fig 1

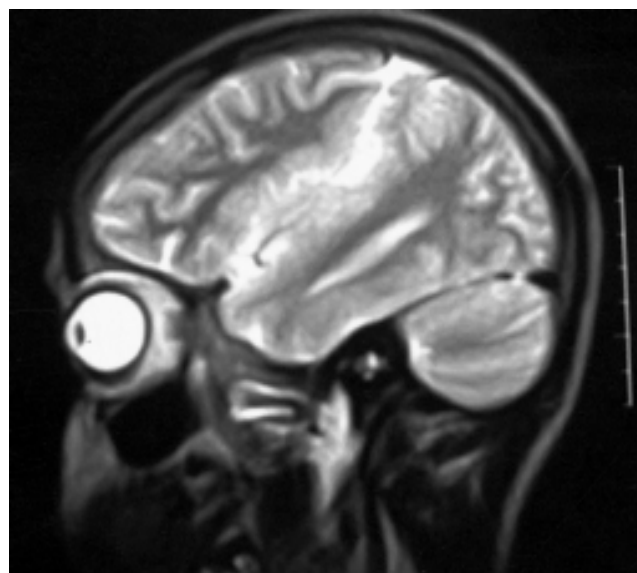


Fig 2

From the Rajiv Gandhi Cancer Institute and Research Center, Sector-5, Rohini Delhi-85

Request for Reprints: Dr. Sangeeta Taneja C-103, Satya Shanti Apartments, Sector-13, Rohini Delhi

Received 20 November 2005; Accepted 15 April 2006

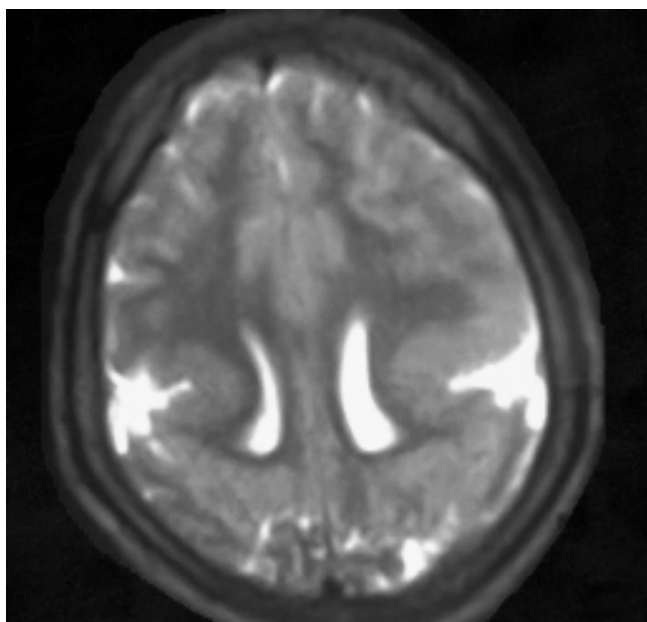


Fig 3

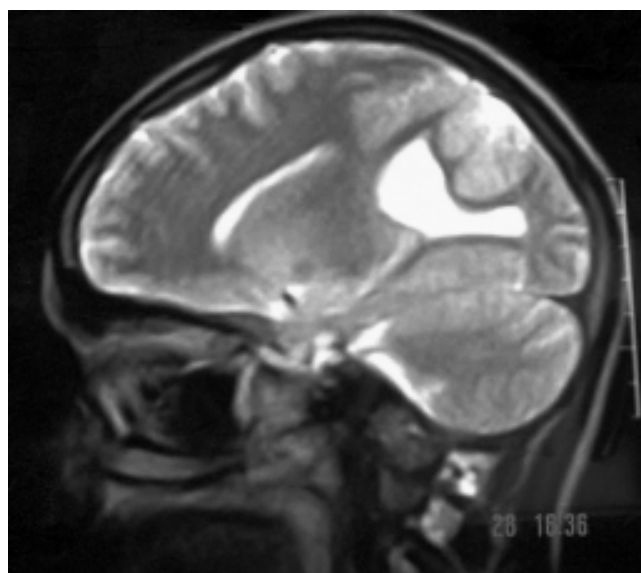


Fig 4

## DISCUSSION

Congenital Bilateral Perisylvian Syndrome is synonymous with Perisylvian Syndrome, Perisylvian polymicrogyria, Worster Drought Syndrome and Bilateral opercular syndrome.

Perisylvian syndrome refers to a neurological disorder in which a particular area of the brain (called the perisylvian region) develops abnormally and the underlying developmental abnormality is Polymicrogyria.

Polymicrogyria refers to abnormal appearance of the cortex with multiple abnormally small convolutions and too few sulci. It is basically an organization anomaly in which the neurons reach their final destination in the cortex but are distributed abnormally. Gross assessment of the thickness of the cortical surface is due to fusion of the adjacent miniature gyri piled upon one another. Multiple syndromes of region specific bilateral symmetrical polymicrogyria have been reported (1).

This syndrome has been associated with congenital cytomegalovirus infection (2), Aicardi syndrome or can be sporadic. The anomaly usually occurs as a result of post-migration insult during fifth or sixth months of pregnancy (3). A new syndrome of Familial Perisylvian Polymicrogyria has been described. However, no definite inheritance pattern could be attributed to the syndrome and it was regarded as genetically heterogeneous. Reports from different families with multiple affected families suggest different modes of inheritance including X linked, autosomal dominant and autosomal recessive (4). It has been reported in identical twin pregnancies complicated by twin twin transfusion syndrome (5). In other cases, it may be due to mutation in a gene.

Syndromes of Bilateral Symmetrical Polymicrogyria have been classified on the basis of their predominant distribution. The presentation of the patient depends on the distribution. Bilateral Frontal Polymicrogyria typically results in developmental delay, mild spastic quadriplegia, variably impaired language development and epilepsy. Bilateral Parasagittal parieto-occipital polymicrogyria is associated with seizures and mild mental retardation; however neurological deficits are often not present. The oromotor dysfunction associated with Bilateral Perisylvian Polymicrogyria is lacking with Bifrontal Malformations

Essential criteria (3) (present in 100% of the cases) for diagnosis of this syndrome are oropharyngoglossal dysfunction, moderate to severe dysarthria and bilateral perisylvian malformations on imaging. Additional criteria (present in more than 85% of the cases) include delayed milestones, epilepsy, mental retardation and abnormal EEG. Other criteria (< 50% of the cases) for diagnosis are arthrogryposis multiplex, other limb malformations and infantile spasms

In our case the patient had all the features except dysphagia, absent gag reflex and arthrogryposis.

Identification of prominent pseudobulbar signs should be sufficient to make a tentative clinical diagnosis, which may be confirmed by imaging. Seizures usually begin between the ages of four and twelve years and are poorly controlled in about 60% of the patients. The most frequent seizure types are atypical absences, tonic or atonic drop attacks and tonic clonic seizures often occurring as

Lennox Gestaut syndrome. A minority of patients have partial seizures (26%).

Polymicrogyria may be difficult to demonstrate with CT but is identifiable on MRI as thickened cortex, poorly developed sulci and an irregular margin at the cortical white matter junction. Abnormalities of cortical venous drainage and DVA's are often present. In Bilateral Perisylvian Polymicrogyria, the opercula are dysplastic and incomplete. The sylvian fissures are wide and underdeveloped. Sagittal Images may show posterior extension of the sylvian fissure, exposure of the insula and apparent thickening of the cortex. The bodies of the lateral ventricles show inverted appearance, typical of this disorder. Proton MR Spectroscopy of the brain allows noninvasive in vivo assessment of metabolites, which may be useful in understanding the biology of malformations of cortical maldevelopment. The neurons of glia in these areas and the metabolites appear to be similar to those of normal adult frontal white matter (6).

Prenatal diagnosis using fetal ultrasound and MRI may be particularly difficult as the regions of the brain that are involved in this malformation may not have reached the final folding until birth. However there have been studies in which patients with Bilateral Polymicrogyria were identified by prenatal MR imaging and genetic analysis was performed (7).

Recognition of the disorder enables appropriate management (3). Because of the severe dysarthria, many

patients are labeled as severely retarded, though they may have normal comprehension. In patients with epilepsy, appropriate but aggressive treatment should be instituted for two reasons. First despite the fixed nature of perisylvian malformations, frequent seizures may exacerbate speech dysfunction. Second, some patients have progressive deterioration, probably related to their epilepsy. In patients with severe and disabling seizures, especially drop attacks, section of the corpus callosum should be considered.

#### REFERENCES:

1. Guerrini R, Barkowich AJ, Hevner R. Syndromes of Bilateral symmetrical Polymicrogyria. *AJNR* 1999; 20:1814-1821.
2. Zucca C. et al Retrospective diagnosis of congenital cytomegalovirus infection and cortical maldevelopment. *Neurology* 2003; 61:710-712.
3. Kuzniecky R, Andermann F, Guerrini R, et al. Congenital Bilateral Perisylvian Syndrome- study of 31 patients. *Lancet* 1993; 341:608-612.
4. Guerreiro M.M., Andermann E, Guerrini R, et al. Familial perisylvian polymicrogyria: A new familial syndrome of cortical maldevelopment *Annals of Neurology* 2000;48:39-48.
5. Barth PG, Vander Harten JJ. Parabolic twin Syndrome with topical isocortical disruption and gastroschisis. *Acta Neuropathol* 1985; 67:345-349.
6. Widjaja Elysa et al. Proton MR Spectroscopy of Polymicrogyria and heterotopia. *AJNR* 2003; 24:2077-2081.
7. Andrea Righini et al. Early Prenatal MR imaging diagnosis of polymicrogyria *AJNR* Feb 2004; 25:343-346.

Imaging Product