

## Case Report

# Congenital bilateral perisylvian syndrome: a rare cause of epilepsy

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### ABSTRACT

Congenital bilateral perisylvian syndrome (CBPS) is an entity proven or diagnosed on basis of neuroimaging in the form of MRI (Magnetic Resonance Imaging). A case with classical triad of fascio-masticatory diplegia, epilepsy and mental retardation has been seen. We report a case of a 6 years old male child on intermittent treatment for seizures since 6 months presenting to us with abnormal movement, recurrent spitting and drooling of saliva with slurring of speech. Diagnosed on MRI with involvement of perisylvian location in temporal lobe. The etiology of epilepsy was justified and symptomatically treated.

**Keywords:** Congenital bilateral perisylvian syndrome, MRI, Epilepsy, Speech

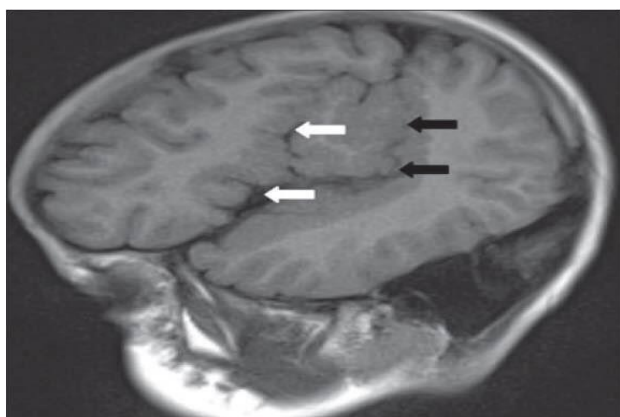
### INTRODUCTION

Congenital bilateral perisylvian syndrome was described in identical twins by Graff – Rafdford, et al. In literature till now only forty patients have been reported.<sup>1</sup> The underlying anomaly in this syndrome is Polymicrogyria which can be focal or regionally distributed or can even involve the whole cortical mantle. The characteristic presentation is epilepsy with associated features involving the fascio-masticatory area presenting as difficulty in swallowing, drooling saliva or speech abnormalities. Mental retardation or minimal cognitive impairment can be also be seen as a part of this syndrome.

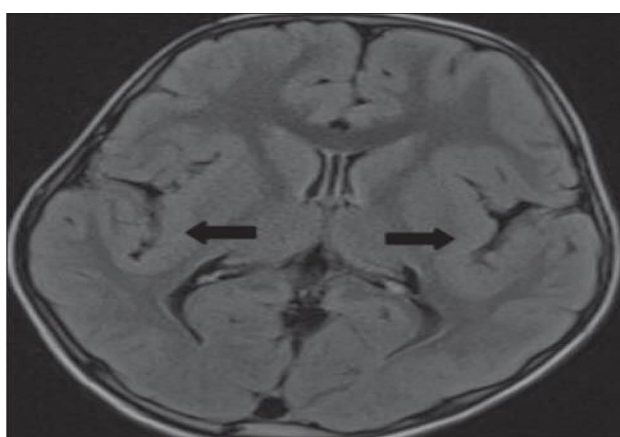
### CASE REPORT

A 19-year-old gravida 1 para 1 woman delivered 6 years old Muslim boy presented with a chief complaint of excessive spitting and shrugging of shoulder. As stated by the mother the frequency of these events was innumerable and had not much significant aggravating or relieving factors. Although if child was made conscious of these movements, it stopped for some time. The history dated back to 6 months when child had 1<sup>st</sup> episode

of generalized tonic seizures and investigations diagnosed with neurocystercosis & treated accordingly; the patient stopped antiepileptic drugs themselves after 3 months. On presentation to our hospital the vitals of the child were stable; Birth history and developmental history was not significant. Anthropometry of the child was within centiles as per the age. CNS examination revealed that the motor and sensory system was intact. Emphasized on the movement of tongue and jaw, it was normal; movements in the form of shrugging of shoulder was seen and assessed. No cranial nerve involvement was appreciated. However on assessing speech there was slurring and dysarthria present. Child was restricted to spit and observed that there was pooling and drooling of saliva. ENT examination showed no significant findings. Child was put on oral antiepileptic as advised after 1<sup>st</sup> episode and was subjected for MRI – Brain that revealed polygyria involving the entire perisylvian cortex with thickening of the grey matter around the sylvian fissure and its widening with irregular contour of the cortex, involving mainly the temporal area; suggestive of Bilateral Perisylvian Syndrome.



**Figure 1: Polymicrogyria involving the perisylvian cortex.**



**Figure 2: Thickening of grey matter around the sylvian fissure with irregular cortex.**

## DISCUSSION

Perisylvian Syndrome refers to a neurological disorder in which a particular area of brain (called the perisylvian region) develops abnormally and the underlying developmental abnormality is polymicrogyria.<sup>2</sup> Polymicrogyria is excessive number of small convolutions (gyri) on the surface of the brain; it can be generalized or focal.

Essential criteria for the diagnosis of this syndrome are oropharyngoglossal dysfunction, moderate to severe dysarthria and bilateral perisylvian malformation.<sup>3</sup> Similar in our case, child has repeated episodes of pooling of saliva, spitting, drooling and difficulty in swallowing. Child also has slurring of speech.

Epileptic seizures are present in majority of cases with CBPS, with broad spectrum of seizure types, including generalized tonic-clonic (GTC), typical and atypical absence, atonic and tonic seizures or drop attack.<sup>3,4</sup> In our case the child has seizures involving deviation of mouth, eye up rolling and frothing from mouth. There is also complaint of excessive shrugging of shoulder. Donders, et al reported that individuals with CBPS may present

with heterogenous psychosocial presentation and that cognitive impairment may not be as prevalent as previously suggested.<sup>5</sup> The child in our space had no cognitive impairment or any signs of mental retardation.

CBPS is supposed to be a migration disorder of the brain associated with distinctive clinical and imaging features.<sup>6</sup> Possible causes known or postulated for this syndrome are injury during neuronal migration, post-migrational vascular accident and gene mutation. The cause in our case was not well defined and the absence of a definitive preceding perinatal insult in the clinical history and MRI changes was against a diagnosis of cerebral palsy.

The best modality for diagnosis is imaging in form of MRI. There has been a study in which patient with bilateral polymicrogyria were identified by prenatal MR imaging and genetic analysis was performed.<sup>7</sup> Different modes of inheritance including X-Linked, autosomal dominant and autosomal recessive from different families have been reported and the mode of transmission remains unknown.<sup>2,3,5,8</sup>

Treatment mainly involves control of seizures with anti-epileptic drugs for long term. Prognosis for epilepsy cannot be predicted based on the early response to treatment.<sup>9</sup>

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