**International Journal of Medical and Health Research**
**ISSN: 2454-9142**
**www.medicalsjournals.com**
**Volume 1; Issue 3; October 2015; Page No. 29-31**

# Bilateral giant open lip schizencephaly with massive subdural effusion

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**Abstract**
Background- Schizencephaly is a rare disorder of neuronal migration which is characterized by clefts in cerebral mantle which communicate between subarachnoid spaces laterally and ventricular system medially.
Case report- An extremely rare case of bilateral giant open lip schizencephaly with massive subdural effusion is presented here in with relevant literature.
Conclusion- There are no guidelines for the treatment of schizencephaly; overall prognosis of schizencephaly remains guarded and depends upon the amount of preserved cortical mantle and associated anomalies.

**Keywords:** Bilateral, Schizencephaly, Subdural hygroma

## Introduction

Schizencephaly is an uncommon disorder of cortical development characterized by a transcerebral cleft, which is lined by gray matter. The clefts extend across the entire cerebral hemisphere, from the ependymal lining of the lateral ventricles to the outer pial surface of the cortex. The clefts are divided into those with fused lips (closed-lips) or type I and those with separated lips (open-lips) or type II schizencephaly. In closed-lips schizencephaly, the walls appose one another directly, obliterating the cerebrospinal (CSF) space within the cleft at that point. In open-lips schizencephaly, CSF fills the cleft all the way from the lateral ventricle to the subarachnoid spaces surrounding the hemisphere[4]. The cavity formed in type II varies in size from small to large ones. There are isolated case reports on both the varities of schizencephaly; though open lip variety with massive subdural hygromas is rarely described in literature. We report a rare case of open lip schizencephaly with bilateral massive subdural hygromas.

## Case Report

A 15 month old female child was brought to our hospital with chief complaints of seizures and developmental delay. Patient was second issue of the couple and was born by a normal vaginal delivery at full term. No antenatal ultrasound was done and patient was not a booked case. Patient remained alright till 2 months until parents noticed an increasing head size. Social smile was delayed and there was no neck holding till 10 months of age. Patient started having generalized tonic clonic seizures at 11 months of age following which he was shown to a pediatrician and CT brain was done. On examination patients head circumference was 50 cm and neck holding and social smile was absent. Limb movement was normal on both sides and there was no sun setting sign. All the primitive release signs were present. CT Brain was showing giant clefts resulting in the connection of lateral ventricles with subarachnoid spaces in both cerebral hemispheres, and interpreted as bilateral giant open-lip schizencephaly; associated with massive bilateral subdural hygromas. The cortical mantle was pushed medially to the extent that the upper cuts of CT film were deficient of any neural structure. MRI Brain confirmed the findings of CT scan. Patient's family was counseled properly and was given the option of Subdural-Peritoneal shunt, which they refused. Patient was started on anticonvulsant medication and seizures were controlled.

## Discussion

In 1946, Yakovlev and Wadsworth first described schizencephaly as hemispheric clefts in the region of the primary fissures, infolding of gray matter along the clefts, and associated cerebral malformations, including ventriculomegaly, polymicrogyria, heterotopias, agenesis of the corpus callosum, and absence of the septum pellucidum[3]. The schizencephaly clefts are mostly perisylvian or centrally located. Holoprosencephaly, arachnoid cyst, hydranencephaly, and porencephaly are included in the differential diagnosis of schizencephaly[3]. The gray matter covering the clefts, which is mostly polymicrogyric and rarely dysplastic, helps in the differential diagnosis. Different theories have been described in the etiology of schizencephaly. Barkovich and Norman have hypothesized a vascular etiology[1]. They proposed that the abnormality results from an infarction in an area of the germinal matrix during the seventh week of embryogenesis. One hypothesis is based on vascular compromise during early neuroembryogenesis. Pathologies like infection, metabolic disorders, ischemia, or genetic defects that cause errors in any of the stages of stem cell differentiation, neuronal migration, or cortical organization are also implicated. The etiology is unclear, although a primary malformation secondary to a neuronal migrational anomaly is considered most likely.

Familial cases of schizencephaly have been reported, suggesting a possible genetic origin within a group of neuronal migration disorders. Iannetti reported cases of clefts resulting from cytomegalovirus infection[2]. Therefore, the appearance of schizencephaly is likely secondary to multiple factors, leading to a final common manifestation of abnormal neuronal migration. One should always remember to screen such cases for associated anomalies.

The symptoms of schizencephaly are variable and depend upon the extent of cortex involved and associated CNS anomalies. The spectrum of symptoms can range from seizures and mild hemiparesis in unilateral closed type schizencephaly, to gross developmental anomalies, motor deficits and quadriparesis in open bilateral clefts. MRI is the imaging modality of choice because of its superior differentiation of gray matter and, due its ability image in more than one plane. There are no guidelines for the treatment of schizencephaly; it consists of treatment of seizures, and physiotherapy, rehabilitation and ventriculoperitoneal shunt in cases complicated by hydrocephalus.

Overall prognosis of schizencephaly remains guarded and depends upon the amount of preserved cortical mantle and associated anomalies.

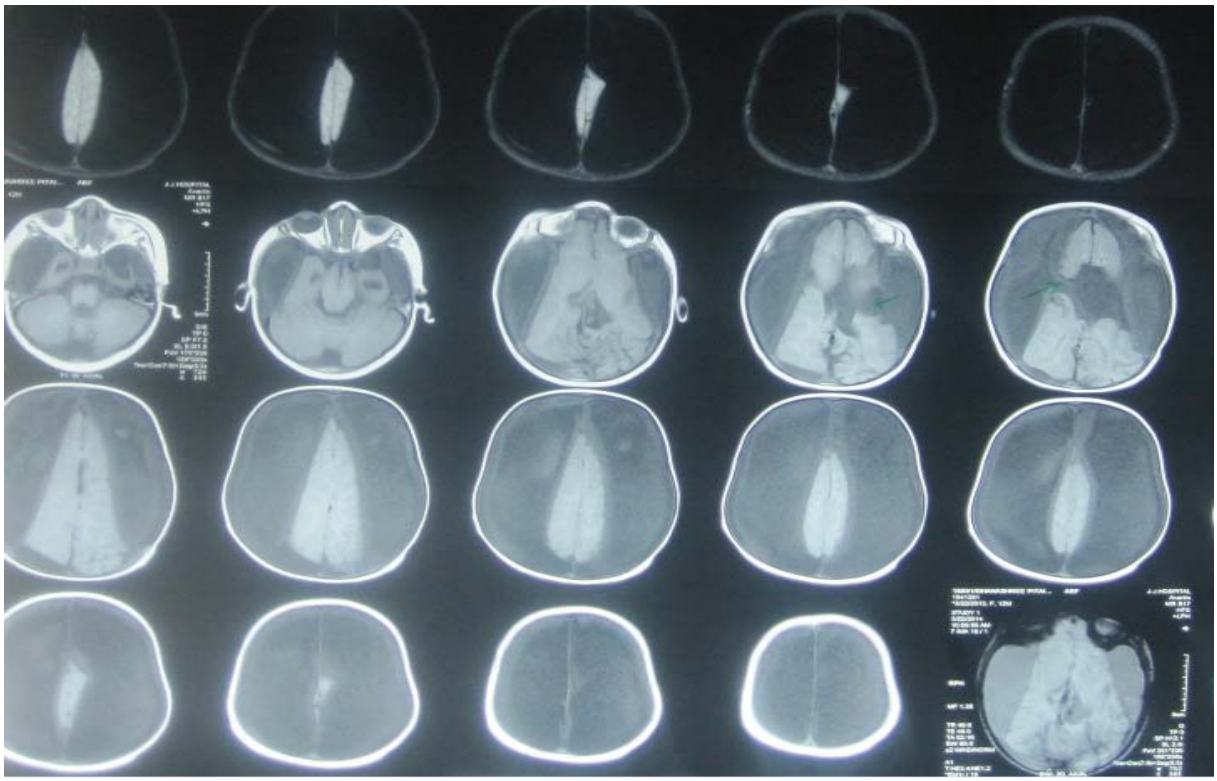

**Fig 1:** MRI Brain with contrast showing bilateral schizencephaly with intervening brain tissue in midline

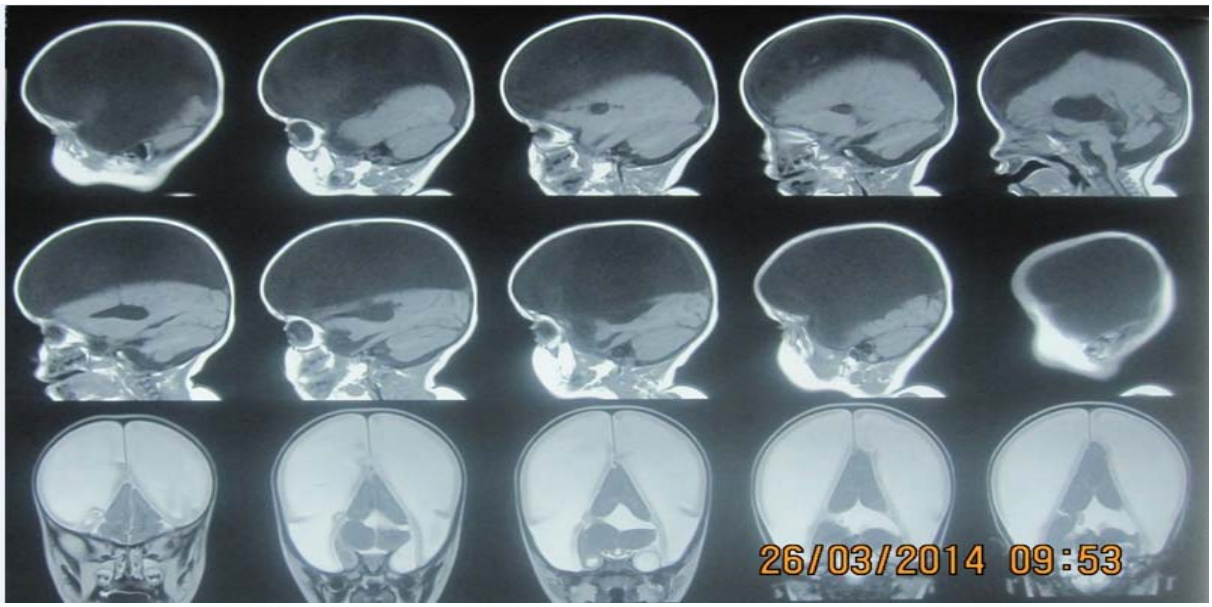


**Fig 2:** MRI Brain with contrast showing bilateral schizencephaly with intervening brain tissue in midline

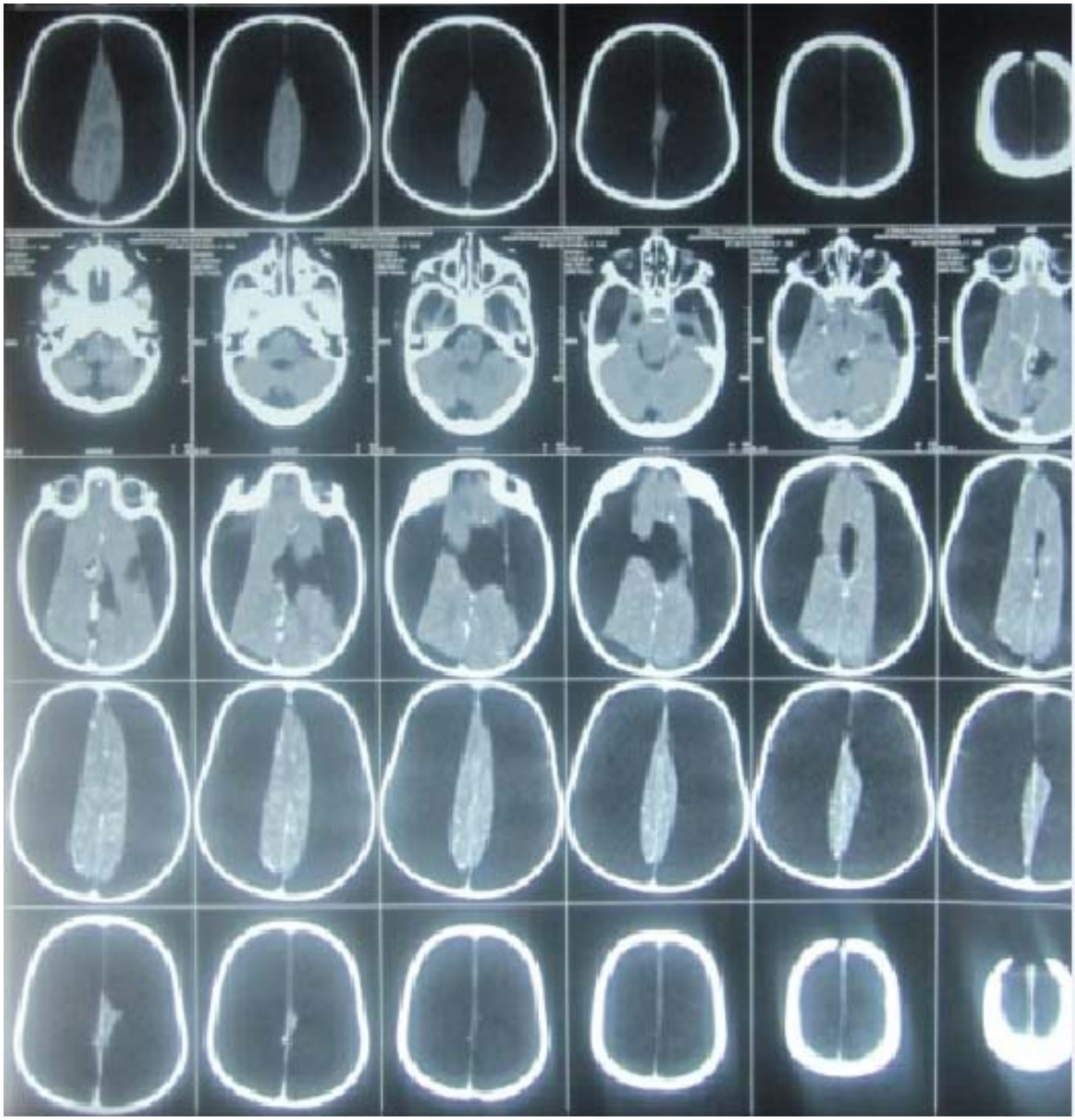

**Fig 3:** CT Brain with contrast of same patient

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