

# Schizencephaly Without Cognitive Deficits: A Case Report

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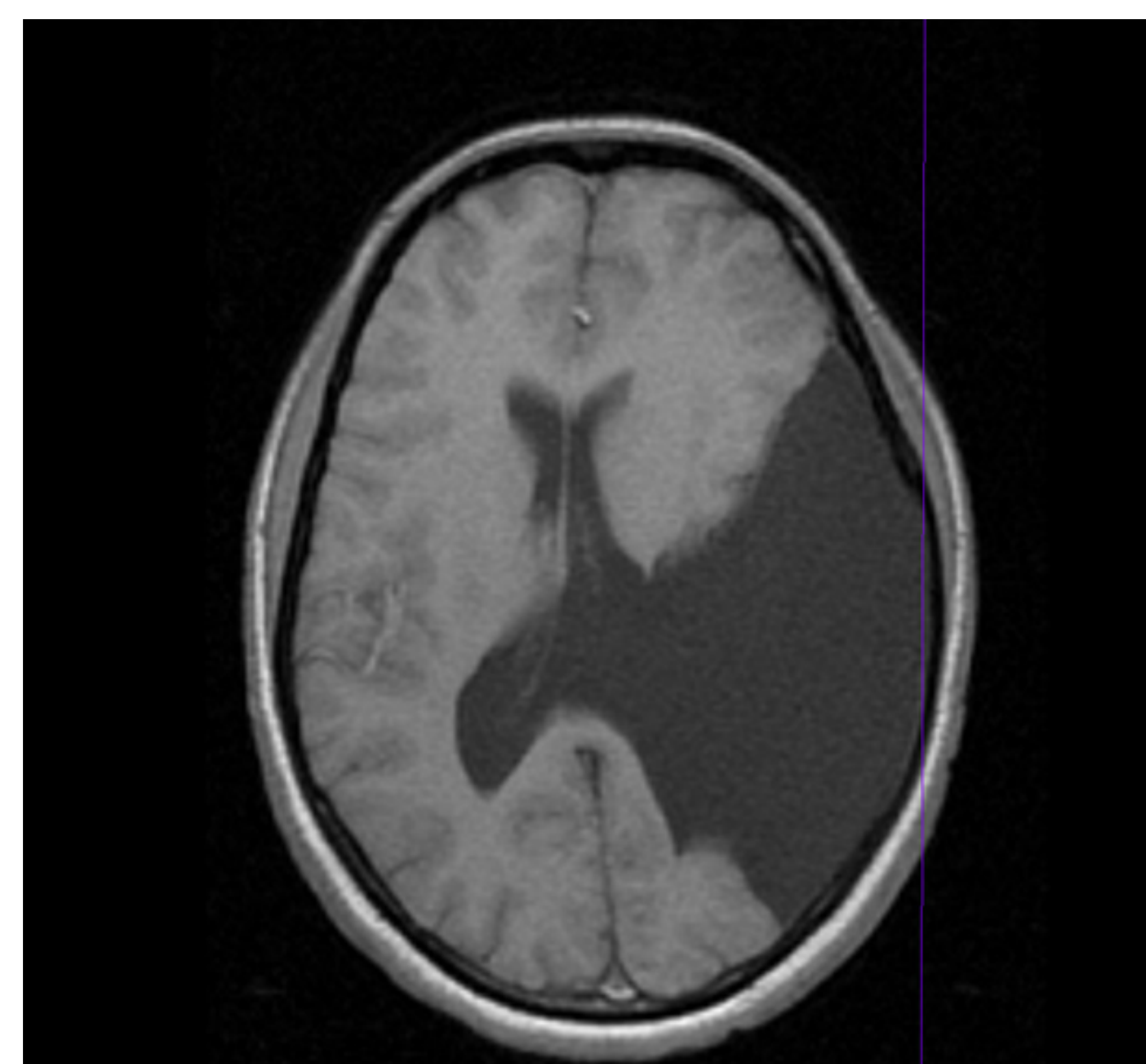


## Setting

Tertiary care teaching hospital

## Case Description

A 17-year-old female presents with right-sided weakness. Prior MRI imaging of the brain showed left-sided open lip schizencephaly with an absence of the posterior aspect of the septum pellucidum. The mother reports that the patient reached all her milestones on time and had no developmental delays. She currently has a 3.6 GPA in high-school and aspires to be a nurse. Upon evaluation, the patient shows no cognitive deficits but has right spastic hemiplegia and trouble with right foot clearance during ambulation. The patient was fitted for a right ankle-foot orthosis and started on a physical therapy program.



MRI of the brain without contrast showing Left-sided open lip schizencephaly with absence of the posterior aspect of the septum pellucidum.

## Case Diagnosis

Schizencephaly is a rare developmental birth defect that occurs in approximately 1 out of 67,000 live births. It is characterized by atypical slits or clefts in cerebral hemispheres that stretch from the lateral ventricle to the cerebral cortex. Most patients with Schizencephaly have neurological impairments affecting motor (90.0%) and/or cognitive (77.5%) functions. Patients often have developmental delay, speech and language deficits, and/or seizures.

## Discussion

Schizencephaly is not curable and treatment is largely supportive. Patients may require physical therapy/ occupational therapy, and bracing to help improve function, prevent falls, and promote safety awareness. Anti-epileptic medications may be prescribed for seizure prophylaxis. In cases complicated by hydrocephalus, ventriculoperitoneal shunt placement may be considered. Patients who exhibit cognitive deficits may benefit from speech therapy and an Individualized Education Plan (IEP) designed to meet the child's specific needs.

## Conclusion

Schizencephaly is a rare, congenital disease with clinical features that primarily involve the neurological and musculoskeletal systems. Once diagnosed, an interdisciplinary approach involving PT/OT/speech therapy, neurology, physiatry, and neurosurgery should be considered to improve function and mobility, manage and prevent seizures, and monitor disease progression. As evident in this case, children with Schizencephaly may not necessarily present with any cognitive deficits. However, for those who do, an IEP may be helpful.

References:  
Kopyta I, Skrzypek M, Raczkiewicz D, Bojar I, Sarecka-Hujar B. Epilepsy in paediatric patients with schizencephaly. Ann Agric Environ Med. 2020 Jun 19;27(2):279-283. doi: 10.26444/aaem/122796. Epub 2020 Jun 2. PMID: 32588606