

CASE OF THE WEEK

PROFESSOR YASSER METWALLY

CLINICAL PICTURE

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8 years old male patient presented clinically with right sided hemiplegia since birth, Lennox Gastaut syndrome (with a history suggestive of infantile spasm during the first few months of life) and mental subnormality. The vision in the left eye was completely lost since birth and the vision in the right eye was markedly defective. The child showed evidence of growth and developmental delay.

RADIOLOGICAL FINDINGS

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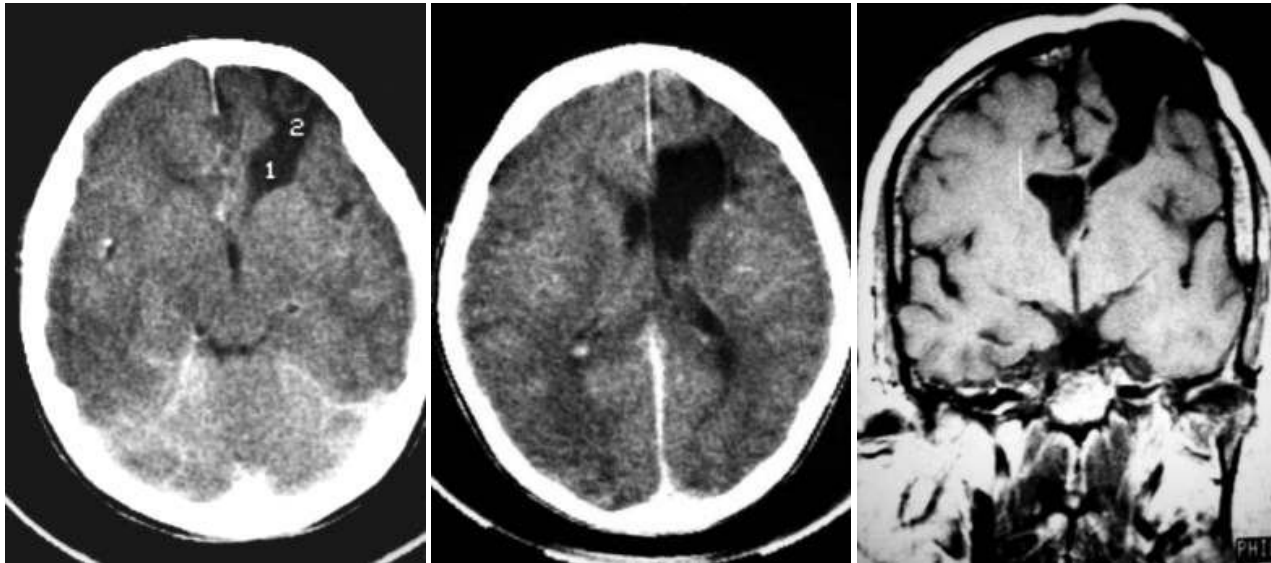


Figure 1. Postcontrast CT scan images (A,B) and Precontrast MRI T1 images (C). Notice the cleft that extends through the entire thickness of cerebral hemisphere from the ventricular surface (ependyma) to the periphery (pial surface) of the brain, apparently the cleft is lined by gray matter (Open lip schizencephaly). The schizencephalic cleft is appreciated at the parasylvian regions and is seen communicating with an encephalocele externally. The optic nerves are markedly hypoplastic bilaterally. The septum pellucidum is markedly deficient, hypoplastic and can not be appreciated in the presented images. The hypoplastic septum pellucidum resulted in a box- like appearance of the frontal horns. It is difficult to see the pituitary stalk in the presented images. The cerebral cortex is lissencephalic and pachygyric and the corpus callosum is markedly thinned anteriorly. The schizencephalic cleft walls are separated (open-lip or type II schizencephaly).

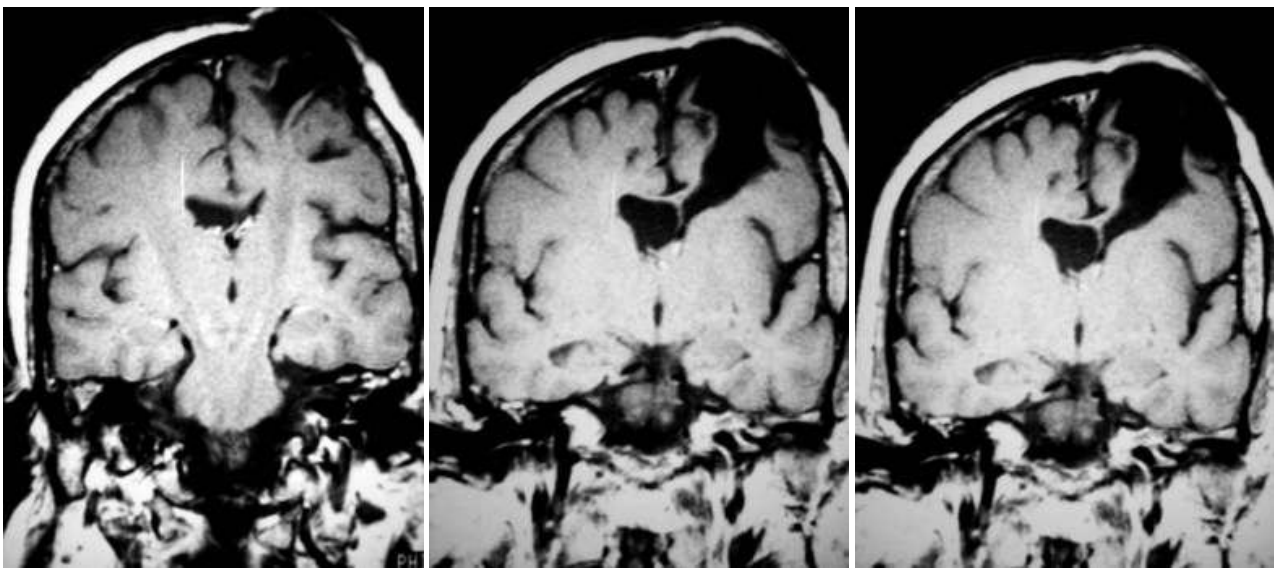


Figure 2. Septo-optic dysplasia. Precontrast MRI T1 images. Notice the cleft that extends through the entire thickness of cerebral hemisphere from the ventricular surface (ependyma) to the periphery (pial surface) of the brain, apparently the cleft is lined by gray matter (Open lip schizencephaly). The schizencephalic cleft is appreciated at the parasylvian regions and is seen communicating with an encephalocele externally. The optic nerves are markedly hypoplastic bilaterally. The septum pellucidum is markedly deficient, hypoplastic and can not be appreciated in the presented images. The hypoplastic septum pellucidum resulted in a box- like appearance of the frontal horns. It is difficult to see the pituitary stalk in the presented images. The cerebral cortex is lissencephalic and pachygyric and the corpus callosum is markedly thinned anteriorly (B,C). The schizencephalic cleft walls are separated (open-lip or type II schizencephaly).

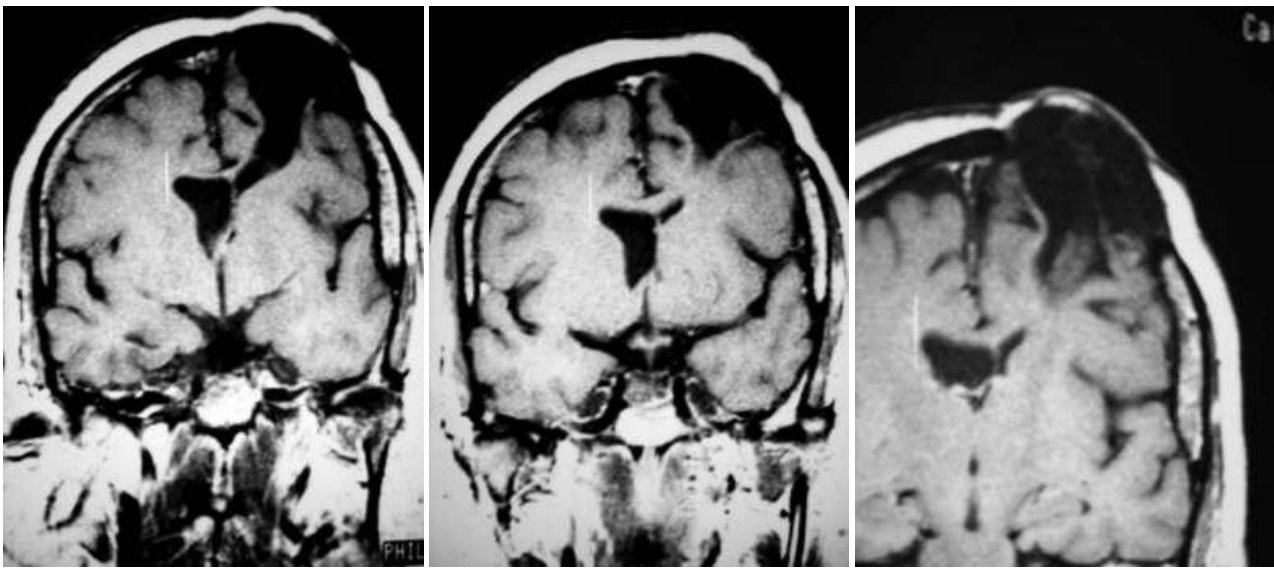


Figure 3. Septo-optic dysplasia. Precontrast MRI T1 images. Notice the cleft that extends through the entire thickness of cerebral hemisphere from the ventricular surface (ependyma) to the periphery (pial surface) of the brain, apparently the cleft is lined by gray matter (Open lip schizencephaly). The schizencephalic cleft is appreciated at the parasylvian regions and is seen communicating with an encephalocele externally. The optic nerves (A) and optic chiasma (B) are markedly hypoplastic bilaterally. The septum pellucidum is markedly deficient, hypoplastic and can not be appreciated in the presented images. The hypoplastic septum pellucidum resulted in a box- like appearance of the frontal horns. It is difficult to see the pituitary stalk in the presented images. The cerebral cortex is lissencephalic and pachygyric and the corpus callosum is markedly thinned anteriorly (A). The schizencephalic cleft walls are separated (open-lip or type II schizencephaly). The pituitary gland appeared vacuolated (B)

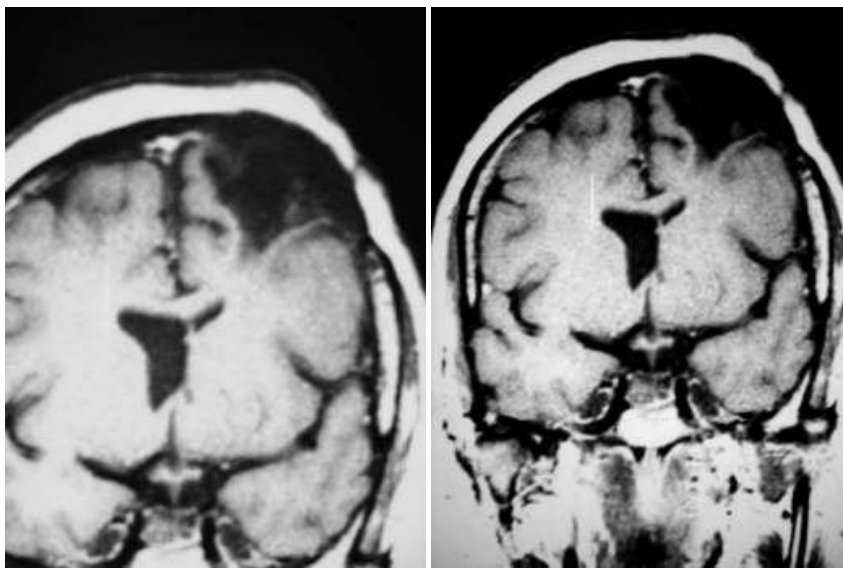


Figure 4. Septo-optic dysplasia. Precontrast MRI T1 images. Notice the cleft that extends through the entire thickness of cerebral hemisphere from the ventricular surface (ependyma) to the periphery (pial surface) of the brain, apparently the cleft is lined by gray matter (Open lip schizencephaly). The schizencephalic cleft is appreciated at the parasylvian regions and is seen communicating with an encephalocele externally. The optic nerves (A) and optic chiasma (B) are markedly hypoplastic bilaterally. The septum pellucidum is markedly deficient, hypoplastic and can not be appreciated in the presented images. The hypoplastic septum pellucidum resulted in a box- like appearance of the frontal horns. It is difficult to see the pituitary stalk in the presented images. The cerebral cortex is lissencephalic and pachygyric. The schizencephalic cleft walls are separated (open-lip or type II schizencephaly). The pituitary gland appeared vacuolated (B)

The encephalocele and the skull wall expansion that are present at the opening of the open-lip schizencephaly (in the presented case) is believed to result from CSF pulsations from the lateral ventricles transmitted through the cleft. [45]

Case summary

- Open-lip or type II schizencephaly
- Deficient and hypoplastic septum pellucidum
- Dysplastic cerebral cortex (The cerebral cortex is lissencephalic and pachygyric)
- The pituitary gland appeared vacuolated
- Hypoplastic optic nerves and optic chiasma
- Hypoplastic corpus callosum
- Encephalocele

DIAGNOSIS:

DIAGNOSIS: SEPTO-OPTIC DYSPLASIA WITH OPEN LIP SCHIZENCEPHALY

DISCUSSION

DISCUSSION:

- **Septo-optic dysplasia (de Morsier syndrome)**

Septo-optic dysplasia (de Morsier syndrome) is a disorder characterized by the absence of the septum pellucidum, optic nerve hypoplasia, and hypothalamic dysfunction. It may be associated with agenesis of the corpus callosum. This disorder should be considered in any patient who exhibits at least two of the above abnormalities and perhaps even solely hypothalamic dysfunction [2]. Septo-optic dysplasia also appears to involve prosencephalic cleavage and development of anterior telencephalic structures [3]. About 50% of patients with septo-optic dysplasia have schizencephaly [1].

Patients may present with visual disturbance, seizures, mental retardation, hemiparesis (especially if associated with schizencephaly), quadriparesis, or hypothalamic dysfunction. Endocrine abnormalities may include growth hormone, thyroid hormone, or antidiuretic hormone function or levels. The consideration of septo-optic dysplasia necessitates an evaluation of the hypothalamic-pituitary axis because as many as 60% of the children with this disorder might exhibit

evidence of a disturbance of endocrine function [4]. This evaluation can include thyroid function studies and electrolytes; these patients are at high risk for growth retardation.

The recent identification of patients with this condition that harbor mutations in the transcriptional regulator gene HESX1, suggest that the mechanism of this disorder is likely genetic and a patterning or segmental abnormality [5]. Even though the genetic abnormality has been identified for a minority of patients, there exists the possibility that this may not represent an entirely genetic disorder because associations have been made with young maternal age, diabetes, the use of anticonvulsants, phencyclidine, cocaine, and alcohol [6,45]

○ Pathology of septo-optic dysplasia

Several congenital malformations might be associated with macroscopic malformations of the septal nuclei. Most of them involve the midline structures, including the septum pellucidum. For example, septo-optic dysplasia is a syndrome characterized by the absence of the septum pellucidum and optic nerve hypoplasia. It may also be associated with pituitary insufficiency resulting in growth and developmental delay. The septal nuclei abnormalities are often an element of a larger cerebral malformation, such as holoprosencephaly, schizencephaly, or pencephaly. In most cases, other midline structures such as the optic nerves, hypothalamus, and septum pellucidum are involved. Absence of the septum pellucidum may reach 80% to 90% in conditions such as schizencephaly. Focal heterotopias or cortical dysplasias may be associated with epileptic seizures. Severe mental retardation and intractable seizures are typical features of the more common larger cerebral malformations and of the syndromes associated with chromosomal anomalies. In some cases of corpus callosum agenesis, it is possible to discern a few axon bundles in an anteroposterior orientation that attempted to cross posteriorly. These are called Probst's bundles. If the corpus callosum is absent, the subcallosal interhemispheric tracts such as the anterior, posterior, and hippocampal commissure might be enlarged. The presence of Probst's bundles is considered by some to indicate that the agenesis of the corpus callosum is due to a primary cause and not the result of secondary injury such as hypoxia. These primary causes include chromosomal abnormalities, single gene defects, and metabolic defects. Cysts and lipomas may be associated with absence of the corpus callosum. [45]

Large occiput encephaloceles may include significant parts of the brain, inducing hypoplasia of the anterior and middle cranial fossa. The brain in the encephalocele could show ischemia or polymicrogyria or be normal. The hypoplasia of the anterior and middle cranial fossa may stretch the septal nuclei, fornix, and septum pellucidum, among other structures. In contrast, transsphenoidal and sphenothmoidal encephaloceles may directly involve the septal nuclei because they may be included in the herniated brain. [45]

○ Neuroendocrine disorders in septo-optic dysplasia

Syndromes that may be associated with pituitary-hypothalamic dysfunction include septo-optic dysplasia, Kallmann's syndrome, and empty sella syndrome. In addition, other developmental anomalies may be associated with pituitary-hypothalamic defects, including agenesis of the corpus callosum, holoprosencephaly, and basal cephaloceles. [45]

Septo-optic dysplasia, a disorder of ventral induction, is considered a mild form of holoprosencephaly. It is a disorder of abnormal induction of the midline mesoderm occurring at the same time as the development of the optic vesicles. Failure of differentiation of the mesoderm into the optic stalk results in aplasia of the stalk. [15] Septo-optic dysplasia is characterized by absence or hypoplasia of the septum pellucidum associated with hypoplasia of the optic nerves as first described by DeMorsier. [12] Associated hypothalamic-pituitary dysfunction occurs in two thirds of patients. [17,18,19] Patients may have hypopituitarism that ranges from panhypopituitarism to growth hormone deficiency, thyroid-stimulating hormone deficiency, elevated prolactin, or ACTH or ADH deficiency. [16] Visual symptoms include nystagmus, amblyopia, or hemianopsia. This syndrome may result from in utero injury or genetic abnormalities. [9] Environmental factors implicated are maternal diabetes, quinidine ingestion, anticonvulsants, alcohol or drug abuse, and cytomegalovirus. [17,45] There may be associated anomalies of neuronal migration, such as schizencephaly and gray matter heterotopias.

On imaging, there is absence or hypoplasia of the septum pellucidum resulting in a box-like appearance of the frontal horns. [8,13,14] Hypoplasia of the optic disks is often diagnosed by an ophthalmologist; however, optic nerve, optic tract, or optic chiasm hypoplasia may be seen in 50% of patients on imaging. [8] Other associated abnormalities include absence of the fornix and callosal dysgenesis. [11,45] On MR imaging, three patterns of involvement have been described in septo-optic dysplasia. [8,11,45]

1. One group has neuronal migration anomalies such as schizencephaly and gray matter heterotopias, hypothalamic-pituitary dysfunction, and partial absence of the septum pellucidum. [45]
2. The other group has complete absence of the septum pellucidum with cerebral white matter hypoplasia. This group may be associated with hypoplasia of the genu of the corpus callosum or hypoplasia of the anterior falx cerebri.
3. Another group of these patients with ectopia of the posterior pituitary has also been described. [11]

Other anomalies that may be associated with complete or partial absence of the septum pellucidum include holoprosencephaly, hydranencephaly, basilar cephaloceles, dysgenesis of the corpus callosum, chronic hydrocephalus,

○ **Optic nerve hypoplasia in septo-optic dysplasia**

Optic nerve hypoplasia, a specific clinical entity, is an increasingly common cause of infantile blindness in the United States. It is characterized by a small optic disc surface area and thin optic nerve caused by a congenital nonprogressive decrease in the number of optic nerve axons. Unilateral and bilateral optic nerve hypoplasia occurs with roughly equal frequency. Various pathophysiologic theories have been proposed, including failure of axons to normally develop; a destructive developmental event, such as a vascular insult; exaggerated apoptosis; or a genetic disorder. [45]

All patients with optic nerve hypoplasia show visual field defects; localized defects as well as generalized constriction are seen. The loss of visual acuity is variable depending on the degree of involvement with axons from the macula, the center of sight. [45]

Optic nerve hypoplasia is recognized as part of a spectrum of optic nerve, central nervous system (CNS), and hypothalamic-pituitary axis congenital anomalies [21]. From the ophthalmologist's perspective, optic nerve hypoplasia can be assigned into five sometimes overlapping groups: isolated unilateral or bilateral optic nerve hypoplasia, with absence of the septum pellucidum (septo-optic dysplasia), with posterior pituitary ectopia or absence of the pituitary stalk, with hemispheric migration anomalies, or with intrauterine and/or perinatal hemispheric injuries [22]. Optic nerve hypoplasia is also associated with a variety of ocular and systemic disorders, including aniridia, albinism, maternal diabetes, fetal alcohol syndrome, and other maternal ingestions [23,24,25].

Groups with optic nerve hypoplasia

- Isolated unilateral or bilateral optic nerve hypoplasia
- Optic nerve hypoplasia with absence of the septum pellucidum (septo-optic dysplasia)
- Optic nerve hypoplasia with posterior pituitary ectopia or absence of the pituitary stalk
- Optic nerve hypoplasia with hemispheric migration anomalies
- Optic nerve hypoplasia with intrauterine and/or perinatal hemispheric injuries

○ **Schizencephaly in septo-optic dysplasia**

Schizencephaly is the most complete form of migrational disorders. It is thought to be caused by a complete agenesis of a section of the cerebral tissue, which results in clefts that extend through the entire thickness of cerebral hemisphere. At the margin of the cleft the pial membrane and ependymal lining of the ventricle lie adjacent to each other and form a pial ependymal seam [29] Schizencephaly probably occurs as the end result of a variety of insults occurring at a critical time and in a critical location during brain development. No specific inciting or unusual prenatal events are described. The lesion is most likely related to multiple etiologies including genetic, toxic, metabolic, vascular or infectious disease. Familial cases have been reported. [30] Septooptic dysplasia, also called as De Morsier disease is a syndrome consisting of blindness, hypoplastic optic nerves and absence of septum pellucidum in females. [29] It is known that Schizencephaly and septo-optic dysplasia frequently coexists. [31] Associated anomalies are heterotopias, Dandy Walker malformation, hydrocephalus and polymicrogyria. [28] Cleft may be unilateral or bilateral, symmetrical or asymmetrical. Commonly located near pre/post central gyrus [26,27]] The cleft in Schizencephaly are lined either totally or in part by gray matter and extend from pial surface to ependyma of the lateral ventricle. The clefts can be located anywhere but commonly occur in parasylvian regions. The cavity formed in open lip type varies in size from small to large and may communicate with lateral ventricle. The ventricular system may be enlarged, particularly in-patients with open lip form of Schizencephaly. [30]

Absence of septum pellucidum, dysgenesis of corpus callosum are often associated with open lip Schizencephaly [28,29] Differential diagnosis includes subarachnoid cysts and porencephaly. Using CT diagnosis of Schizencephaly is sometimes difficult particularly in case of type I. CT may show a slight outpouching at the ependymal surface of cleft and a full thickness cleft may be difficult to identify on CT Scan .Secondary findings like hydrocephalus, heterotopia, polymicrogyria, subdural hygromas and arachnoid cysts can be identified. [30] MRI gives most detailed and precise definition of anatomy and anomaly. For anatomy MRI T1 images suffice [28] MRI is the modality of choice. MRI better delineates the gray matter lining the cleft, which is pathognomonic finding in Schizencephaly.MRI also provides superb cortical anatomy detail and multiplanar capacity. Primary findings related to the cleft and secondary findings associated with Schizencephaly are identified using MRI.The ability of MRI pulse sequence to differentiate gray matter and white matter permit demonstration of gray matter heterotopias in the subcortical white matter beneath the cleft, abnormalities involving the cortex (pachygyria and polymicrogyria) and other secondary findings are also identified. Homolateral absence of sylvian vasculature, small medullary pyramids low position of fornix and thinning of the corpus callosum are findings related to absent cerebral cortex and are better demonstrated by MRI than with other studies.

Sonography can be done in neonatal period in-patients in whom this anomaly is suspected. In Schizencephaly type I, a

hyperechoic line extends from the parasylvian region to the anterior portion of lateral ventricle. The hyperechoic line represents the cortex lining the fused cleft. This type of anomaly is difficult to detect with ultrasound and requires high index of suspicion and highly skilled operator. In Schizencephaly type II an anechoic band or cavity, representing the fluid filled cleft extends from the cortical surface of lateral ventricle. The meeting of the closed lip portion or apex of the cleft with the margin of ventricle may be identified as a ventricular diverticulum or dimple. The size of the caudate, thalamus and lenticular nuclei (subcortical gray matter structures) is decreased. Other associated anomalies such as ventricular enlargement may also be identified. [30]

This case represents a typical association of septo-optic dysplasia with open lip Schizencephaly with all cardinal neurological cardinal features on one hand and a rare form of unilateral open lip cleft on the other. It highlights the importance of MRI in diagnosis of such a condition.

SCHIZENCEPHALY

Schizencephaly is an uncommon disorder of neuronal migrational characterized by a cerebrospinal fluid-filled cleft, which is lined by gray matter. The cleft extends across the entire cerebral hemisphere, from the ventricular surface (ependyma) to the periphery (pial surface) of the brain.

The clefts may be unilateral or bilateral and may be closed (fused lips), as in schizencephaly type I, or separated (open lips), as in schizencephaly type II. Presentation and outcome are variable, but patients typically present with seizures, hemiparesis, and developmental deficits. Usually, the severity of symptoms is related to the amount of brain affected by the abnormality.

- **Aetiology and types of schizencephaly**

Several theories have been proposed to explain the etiology of schizencephaly, although none is universally accepted. The leading theory indicates that schizencephaly results from an early, focal destruction of the germinal matrix and surrounding brain before the hemispheres are fully formed. Schizencephaly probably occurs as the end result of a variety of insults occurring at a critical time and in a critical location during brain development. No specific inciting or unusual prenatal events have been identified, and reported cases are sporadic. The lesion is most likely related to multiple etiologies, including genetic, toxic, metabolic, vascular, or infectious causes. Familial cases of schizencephaly have been reported. Schizencephaly is uncommon; to our knowledge, there are no documented geographical differences in its occurrence. Severity of the symptoms depends on the amount of brain involved. Affected patients typically have seizures, hemiparesis, variable developmental delay, and blindness. Patients also have variable degrees of mental retardation. Patients with open-lip schizencephaly die at an earlier age than patients with the closed-lip form. Usually, death results from failure to thrive, chronic infections, and respiratory problems. Patients with closed-lip schizencephaly may not present clinically until later in infancy or early childhood and may live to early adulthood.

Schizencephaly is divided into 2 types, which have prognostic significance. In closed-lip or type I schizencephaly, the cleft walls are in apposition. In open-lip or type II schizencephaly, the cleft walls are separated. Schizencephaly type II occurs more commonly than type I.

The clefts in schizencephaly are lined either totally or in part by gray matter and extend from the pial surface to the ependyma of the lateral ventricle. The clefts can be located anywhere, but they commonly occur in the parasylvian regions. The clefts can be unilateral or bilateral, and can be either symmetric or asymmetric. The cavity formed in the open-lip type varies in size from small to large and may communicate with the lateral ventricle. The ventricular system may be enlarged, particularly in patients with the open-lip form of schizencephaly.

Gray-matter heterotopia (collections of gray matter in abnormal locations), polymicrogyria, and arachnoid cysts can be associated with schizencephaly. Heterotopias and polymicrogyria typically line the clefts. Microcephaly has been noted in some patients. The septum pellucidum is absent in 80-90% of patients, and schizencephaly may coexist with septo-optic dysplasia.

- **Clinical picture of schizencephaly**

Clinical features of schizencephaly are highly variable. Patients with unilateral clefts with fused lips may have mild hemiparesis and seizures but otherwise have normal development. When the cleft is open, patients present with mild-to-moderate developmental delay and hemiparesis; severity is related to the extent of cortex involved in the defect.

Patients with bilateral clefts present with severe mental deficits and severe motor anomalies including spastic quadriplegia. Frequently, these patients present with blindness, which is often associated with optic nerve hypoplasia. Language development is more likely to be normal in patients with unilateral schizencephaly compared to patients with bilateral clefts.

Intractable seizures frequently are noted in schizencephaly. Several types of seizures have been reported including generalized tonic-clonic, partial motor, and sensory seizures. Infantile spasms have been seen in a few children. Reports

have found no correlation between the subtype of schizencephaly and the occurrence or type of seizure disorder. Identification of gray matter lining the cleft is the pathognomonic finding in differentiating schizencephaly from porencephaly; this is best demonstrated on MRIs.

The more complete information obtained by MRI enables a more accurate prediction of neurologic outcome.

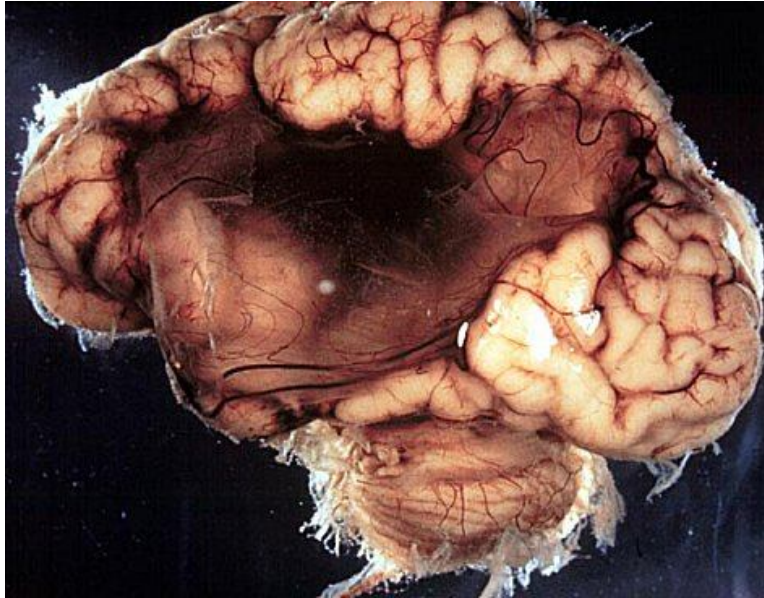


Figure 5. **Open-lip schizencephaly with cortical dysplasia**



Figure 6. **Bilateral open lip schizencephaly**

- **Neuroimaging of schizencephaly**
 - **CT scan**

Using CT, the diagnosis of schizencephaly is sometimes difficult, particularly type I, or closed lip schizencephaly. CT scans of closed-lip schizencephaly may show only a slight outpouching at the ependymal surface of the cleft, and a full-thickness cleft may be difficult to identify on CT scan. The cleft is partially or totally lined by gray matter and extends from the lateral ventricle to the pial surface of the cerebral hemisphere. Secondary findings that can be identified on CT

scan include hydrocephalus, heterotopia, polymicrogyria, subdural hygromas, and arachnoid cysts.

- **MRI**

MRI is the modality of choice for evaluating patients with schizencephaly. MRI better delineates the gray matter lining the cleft, which is the pathognomonic finding in schizencephaly. MRI also provides superb cortical anatomy detail and multiplanar capability. Primary findings related to the cleft and secondary findings associated with schizencephaly are identified using MRI.

The ability of MRI pulse sequences to differentiate gray matter and white matter permits demonstration of gray-matter heterotopias in the subcortical white matter beneath the cleft, abnormalities involving the cortex (eg, pachygyria or polymicrogyria), and other secondary findings also identified by using CT scans. Homolateral absence of the sylvian vasculature, small medullary pyramids, a low position of the fornix, and thinning of the corpus callosum are findings related to absent cerebral cortex and are better demonstrated by MRI than with other studies.

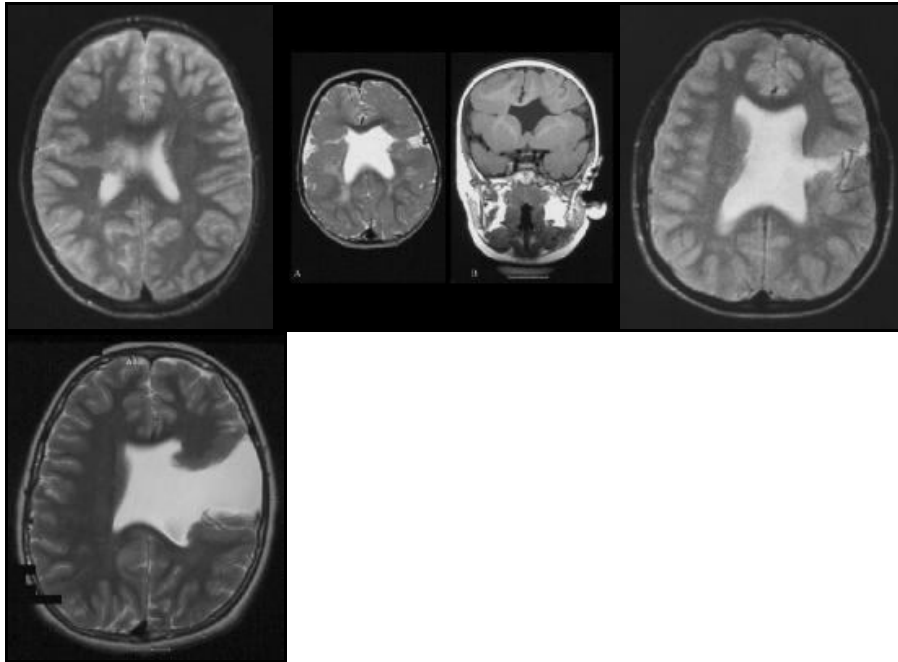


Figure 7. A, Schizencephaly. Axial T2-weighted MRI in unilateral closed-lip (type I) schizencephaly. The cleft is lined by gray matter and extends from the pial surface to the lateral ventricle. B, Schizencephaly. Axial T2-weighted (left) and coronal T1-weighted (right) MRIs in bilateral closed-lip (type I) schizencephaly. A ventricular diverticulum defines the meeting of the closed-lip portion of the clefts with the margin of the ventricles. The septum pellucidum is absent, and the clefts are lined by gray matter and extend from the pial surface to the lateral ventricle. C, Schizencephaly. Axial T2-weighted MRI demonstrates a small open-lip schizencephaly. The septum pellucidum is absent. D, Schizencephaly. Axial T2-weighted MRI in unilateral open-lip (type II) schizencephaly. The septum pellucidum is absent, and a large cerebrospinal fluid–filled cleft extends from the lateral ventricle to the cortical surface. The cleft is lined by gray matter. E, Schizencephaly. Coronal sonograms with a corresponding coronal T1-weighted MRI of open-lip bilateral schizencephaly. Extensive bilateral schizencephalic defects with large CSF-filled clefts extend from the lateral ventricles to the cortical surface.

SUMMARY

SUMMARY

Schizencephaly is the term describing gray matter-lined clefts that extend through the entire hemisphere from the lateral ventricles to the cortical surface, characterized pathologically by a pial-ependymal continuity [28,29]. The gray matter lining these clefts is dysplastic and has irregular inner and outer surfaces, identical to that of polymicrogyria. The clefts can be unilateral or bilateral and are most commonly located near the pre- and postcentral gyri, locations almost

identical to those in which polymicrogyria occurs [22,23]. It is probable that schizencephaly represents an extreme variant of cortical dysplasia, in which the infolding of cortex extends all the way into the lateral ventricle [22,23]. The lips of the cleft may be fused, in which case the walls are directly apposed, obliterating the CSF space within the cleft at that point. When the lips are separated, CSF fills the cleft from the lateral ventricle to the subarachnoid space surrounding the hemispheres [23,28-30]. As in polymicrogyria, large vessels are often seen in the cleft between the lips of the schizencephaly.

The severity of the clinical symptoms is related to the amount of involved brain [23, 31,32]. Patients with a unilateral cleft with fused lips typically present with mild hemiparesis or epilepsy but are otherwise developmentally normal. Patients with unilateral clefts with separated lips more commonly present with hemiparesis and a mild to moderate developmental delay, depending on the location of the cleft within the brain. Patients with bilateral clefts tend to be severely retarded, with refractory seizures beginning at a very early age and severe motor anomalies, and they may appear to be blind. Optic nerve hypoplasia is seen in up to one-third of affected patients [8,30, 33,34].

Routine spin-echo MR obtained in at least two planes is usually adequate for imaging of schizencephalies. These studies show full-thickness clefts, lined by gray matter with an irregular inner surface, that extend through the hemisphere from the ventricle to the surface of the brain. The gray matter lining the cleft often extends into the ventricle as a subependymal heterotopia [6,8,23,30,32]. A dimple is usually seen in the wall of the lateral ventricle where it communicates with the cleft. The dimple provides a helpful sign of continuity of the cleft with the ventricle when the lips of the cleft are fused. The gyral pattern of the cortex adjacent to the cleft is usually abnormal, with features characteristic of polymicrogyria. Polymicrogyria may also be present in the hemisphere contralateral to a unilateral schizencephaly [22,23]. Therefore, the contralateral hemisphere should always be scrutinized.

The calvarium is often expanded over the opening of an open-lip schizencephaly. The expansion is believed to result from CSF pulsations from the lateral ventricles transmitted through the cleft. When plagiocephaly is severe, insertion of a ventriculoperitoneal shunt may help to dampen the pulsations and reverse the cranial asymmetry.

- **Addendum**

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