Case 14721

Eurorad

Fetal MRI of post-clastic schizencephaly

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Section: Neuroradiology
Area of Interest: Foetal imaging Neuroradiology brain
Procedure: Education
Imaging Technique: MR
Imaging Technique: Mammography
Imaging Technique: Experimental
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 25 years, female

Clinical History:

25-year-old female G4P1A2, 29 weeks of gestation with obstetric ultrasound of concern for ventriculomegaly and schizencephaly.

Imaging Findings:

Examination demonstrated fetus in cephalic vertex position. Placenta was along the posterior uterine wall extending towards the fundus. Sidedness of fetal body was established by cardiac configuration and gallbladder position. Fetal brain showed bilateral open lip schizencephaly with expanded lateral ventricles at the schizencephaly defects. Larger left perisylvian schizencephaly defect projected outwards from the body of the left lateral ventricle above the left lenticulate nucleus, best demonstrated on coronal series. Contralateral right frontal schizencephaly was more anterior, centred at the opercular margin radiating from the right frontal horn. There was normal-appearing corpus callosum at midline. Third ventricle and fourth ventricle were normal. There was a megacisterna magna behind a normal-appearing cerebellum. Craniocervical junction, cervical and thoracic spinal cord, conus medullaris and lumbar spine appeared normal. (Figures 1, 2 and 3)

Discussion:

Schizencephaly is an important cause of an abnormal cerebrospinal fluid (CSF) collection in the fetus. Abnormal fetal brain fluid differential diagnosis includes developmental lesions and destructive lesions. Developmental lesions include arachnoid cyst, ventriculomegaly, monoventricule in holoprosencephaly, agenesis of the corpus callosum with an interhemispheric cyst and schizencephaly. Destructive lesions include porencephalic cyst, ventriculomegaly from infection or bleeding and hydranencephaly. [1].

Schizencephaly is a very rare neuronal migration disorder characterized by full thickness clefts within the cerebral hemisphere extending from the hemispheric pial surface to the lateral ventricle ependymal border, being lined by cortical and dysplastic grey matter [2, 3], which differentiates schizencephaly from porencephalic cyst, its main differential diagnosis, which is lined by gliotic white matter, with no grey matter (Fig. 4).

Schizencephaly is classically divided in close-lip (Type 1) and open lip (Type 2). In Type 1 the transmantle column of grey matter is solid without the central channel of CSF. In Type 2 there is a CSF filled cleft from pial surface to the ventricle through the centre of the column of grey matter (Fig. 4). Open lesions have been subclassified in small or large according to size [4]. Schizencephaly can also be subdivided in unilateral or bilateral or cerebral or cerebellar. Schizencephaly is associated with polymicrogyria, cortical dysplasias, dysplasias of the corpus callosum or septum

pellucidum and septo-optic dysplasia.
The cause of this defect is not clear, but some authors believe that intrauterine ischaemia or expressions of genetic factors damage the germinal matrix, with resulting impaired cellular migration at 6–7 weeks of intrauterine life.
Clinical manifestations in unilateral schizencephaly include hemiparesis, mild mental delay, and drug-resistant epilepsy. Bilateral schizencephaly clinical manifestations include spastic tetraplegia, severe mental deficit but less likelihood of drug resistant epilepsy. The size of the malformation does not correlate to the severity of the epilepsy. The prenatal diagnosis is important for patient counseling and pregnancy management.
Fetal MR imaging is considered the study of choice for the evaluation of suspected schizencephaly, with 100% of sensitivity and specificity [5], since it easily delineates grey matter lining the cortical clefts, differentiates Type 1 and Type 2 and detects the associated anomalies. The imaging appearance is identical pre and postnatally.
Functional MR imaging is a tool to identify the eloquent cortex in the planning for epilepsy surgery [6] and the plasticity-related reorganization of brain function later on in childhood.
Differential Diagnosis List: Bilateral schizencephaly, Porencephalic cyst, Focal cortical dysplasia, Heterotopic gray matter, Arachnoid cyst

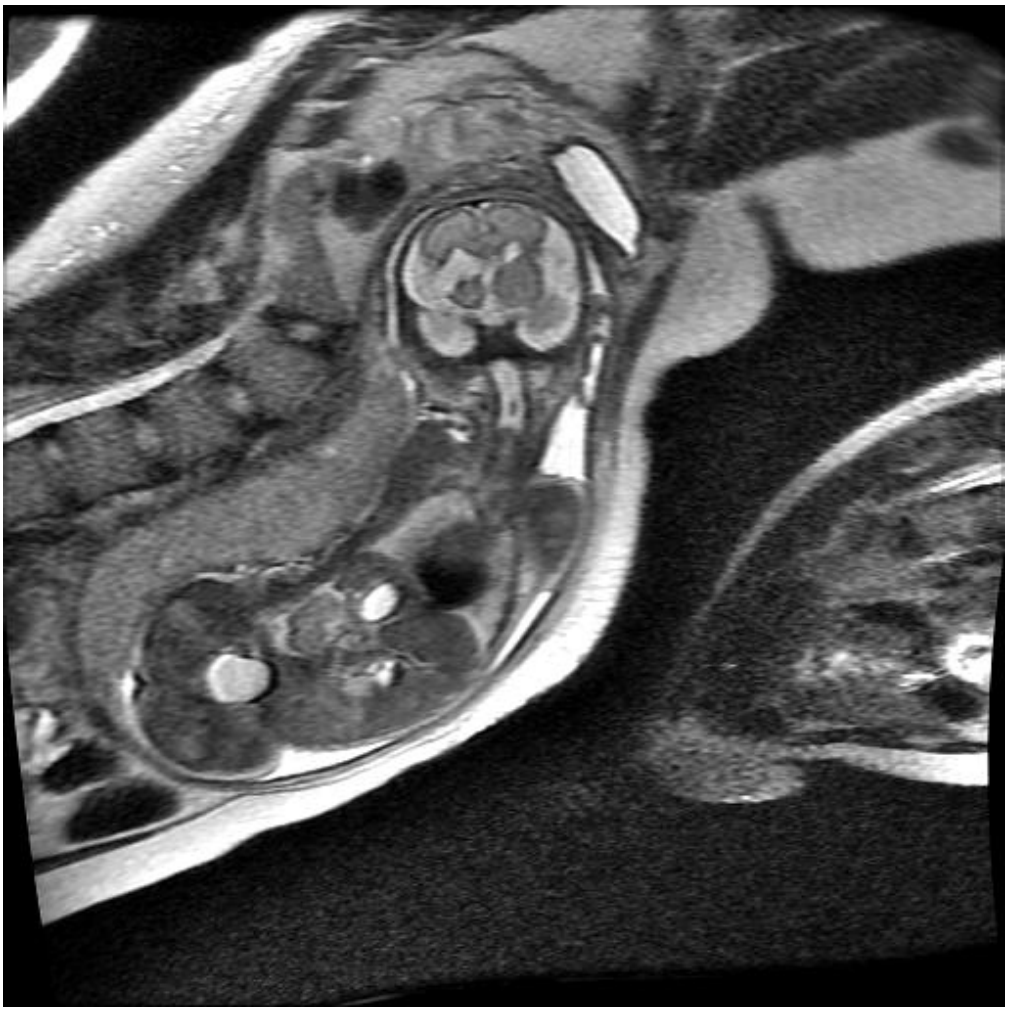
Final Diagnosis: Bilateral schizencephaly

References:

Oh KY, Kennedy AM, Frias AE Jr, Byrne JL. (2005) Fetal schizencephaly: pre- and postnatal imaging with a review of the clinical manifestations. Radiographics May-Jun;25(3):647-57. (PMID: 15888615)
Barkovich AJ, Norman D. (1988) MR imaging of schizencephaly. AJR Am J Roentgeno Jun;150(6):1391-6. (PMID: 3259384)
Denis D, Chateil JF, Brun M, Brissaud O, Lacombe D, Fontan D, Flurin V, Pedespan (2000) Schizencephaly: clinical and imaging features in 30 infantile cases. Brain Dev Dec;22(8):475-83 (PMID: 11111060)
Naidich TP, Griffiths PD, Rosenbloom L. (2015) Central nervous system injury in utero: selected entities. Pediatr Radiol Sep;45 Suppl 3:S454-62 (PMID: 26346151)
Nabavizadeh SA, Zarnow D, Bilaniuk LT, Schwartz ES, Zimmerman RA, Vossough A. (2014) Correlation of prenatal and postnatal MRI findings in schizencephaly. AJNR Am J Neuroradiol Jul;35(7):1418-24 (PMID: 24610904)
Bartolini L, Kao A, Zachery A, Murnick JG, Gaillard WD (2017) Teaching NeuroImages: Homotopic motor distribution on fMRI in closed-lip schizencephaly. Neurology Jan 10;88(2):e24-e25 (PMID: 28069984)

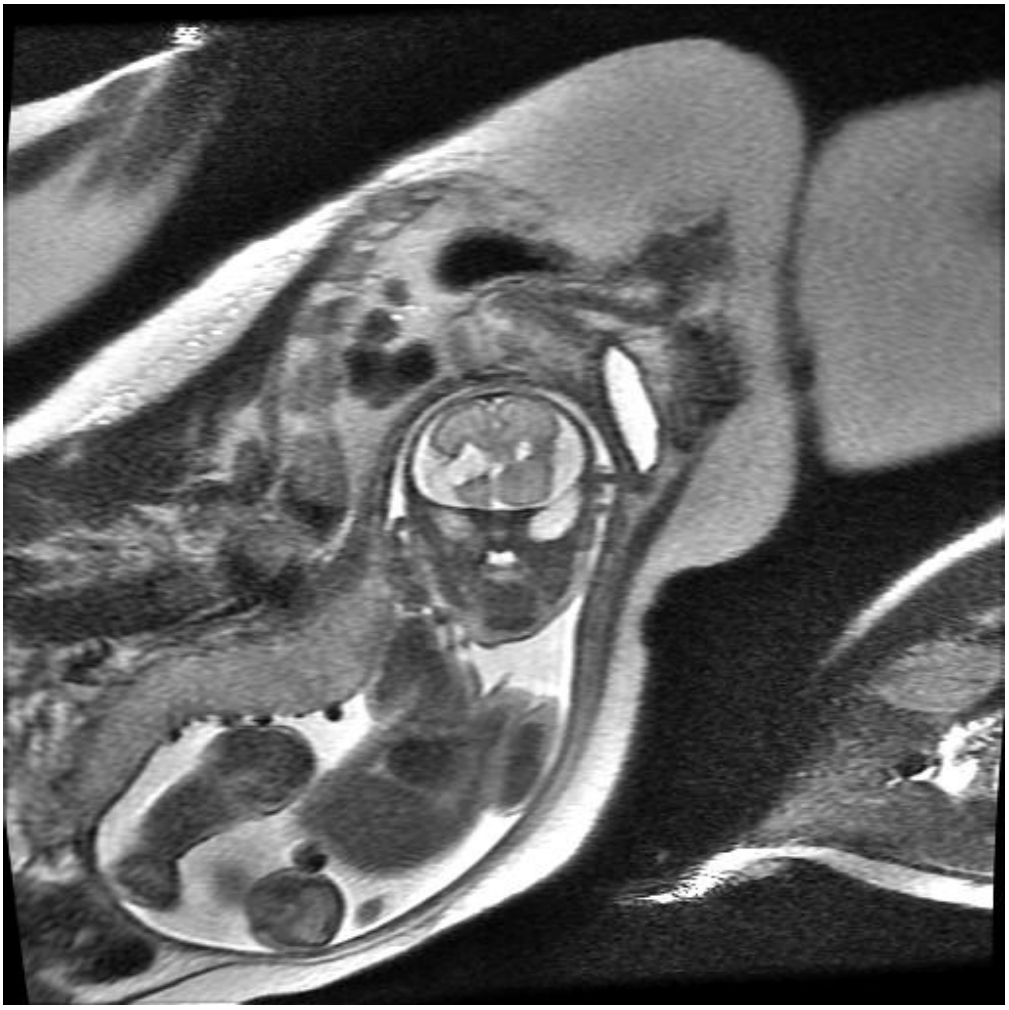
Figure 1

a



Description: Fetal brain shows bilateral open lip schizencephaly with asymmetry of the lateral ventricles at the schizencephaly defects.
T2 hypo rim is visible as sign of post-haemorrhagic tissue disruption. **Origin:** Augusta University

b



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c

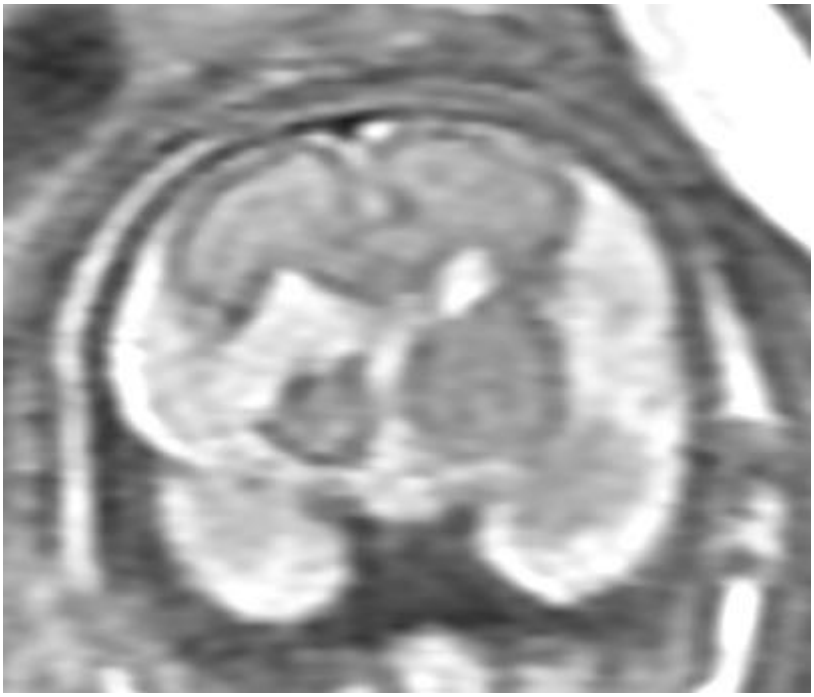

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Figure 2

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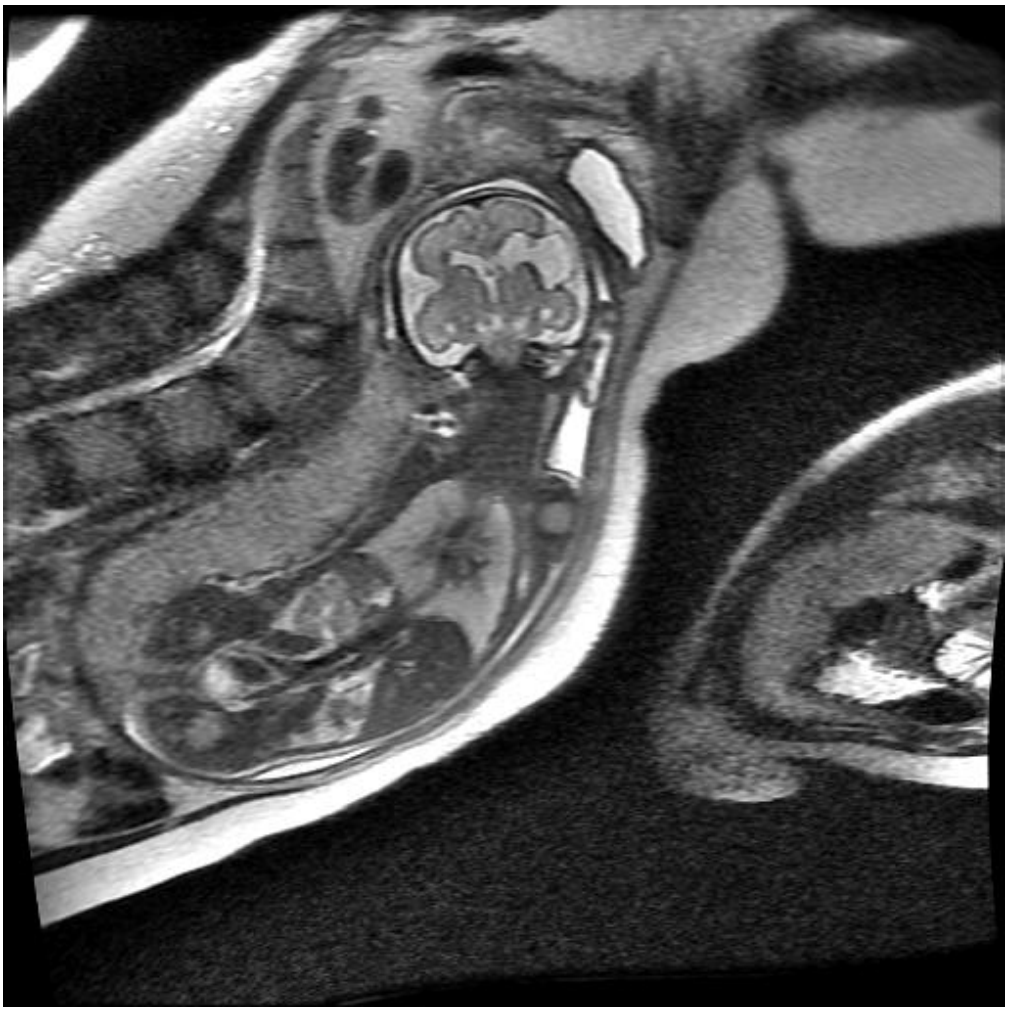

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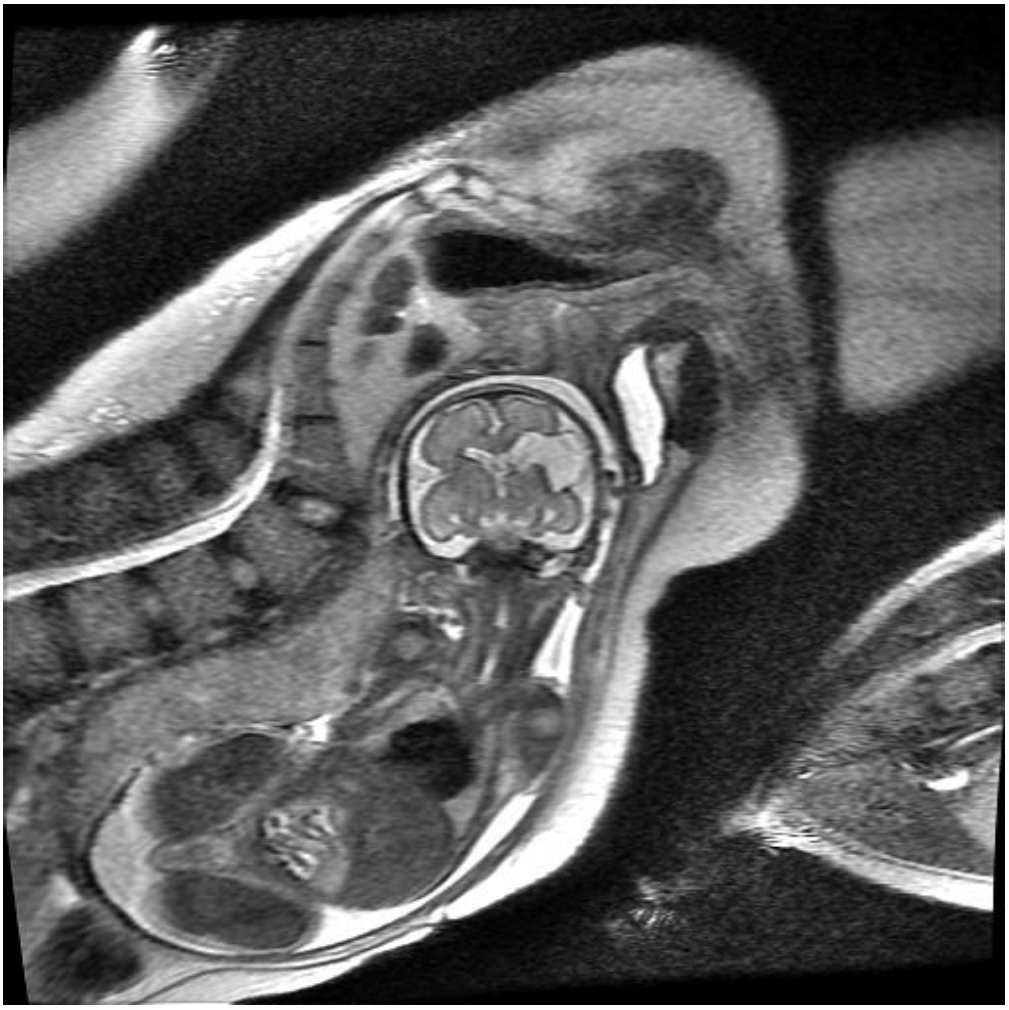

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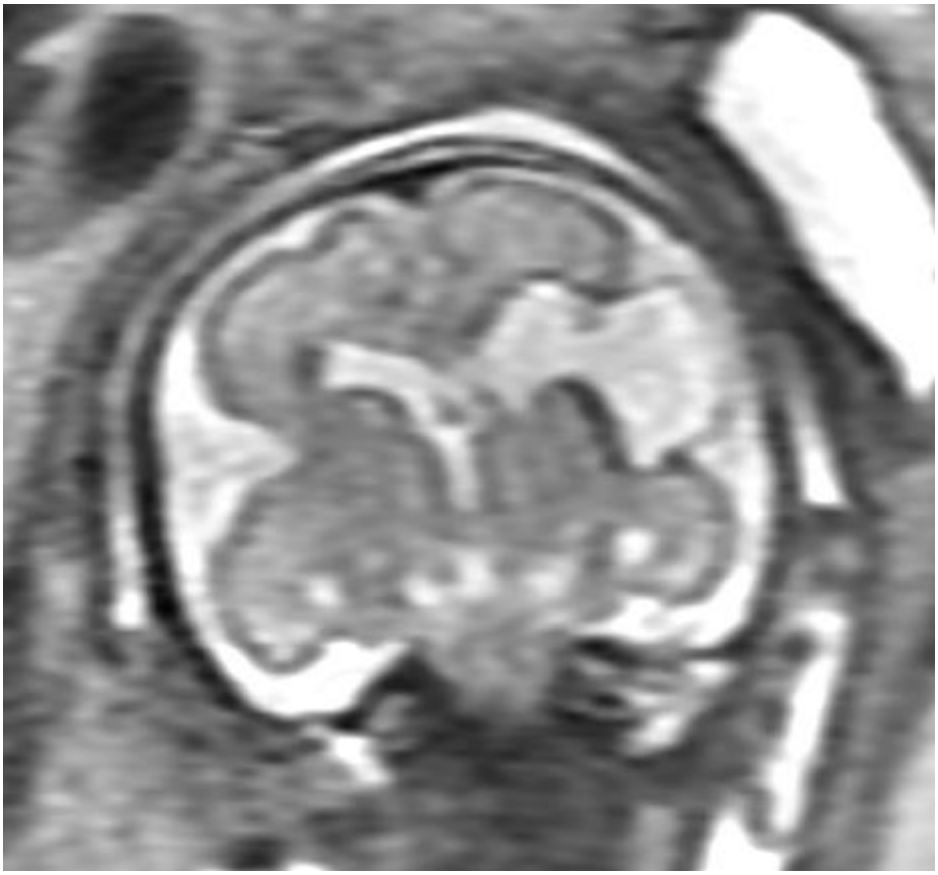
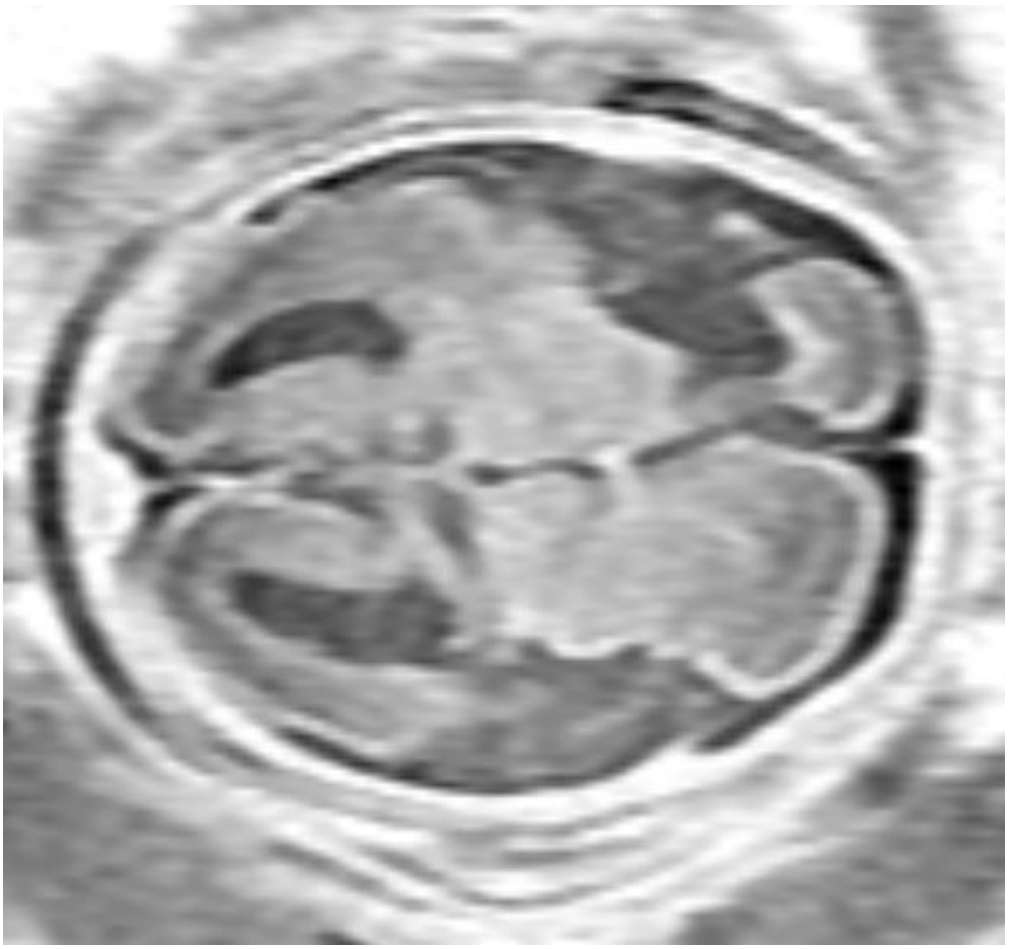
c



Description: Fetal brain shows bilateral open lip schizencephaly with asymmetry of the lateral ventricles at the schizencephaly defects.
T2 hypo rim is visible as sign of post-haemorrhagic tissue disruption. **Origin:** Augusta University

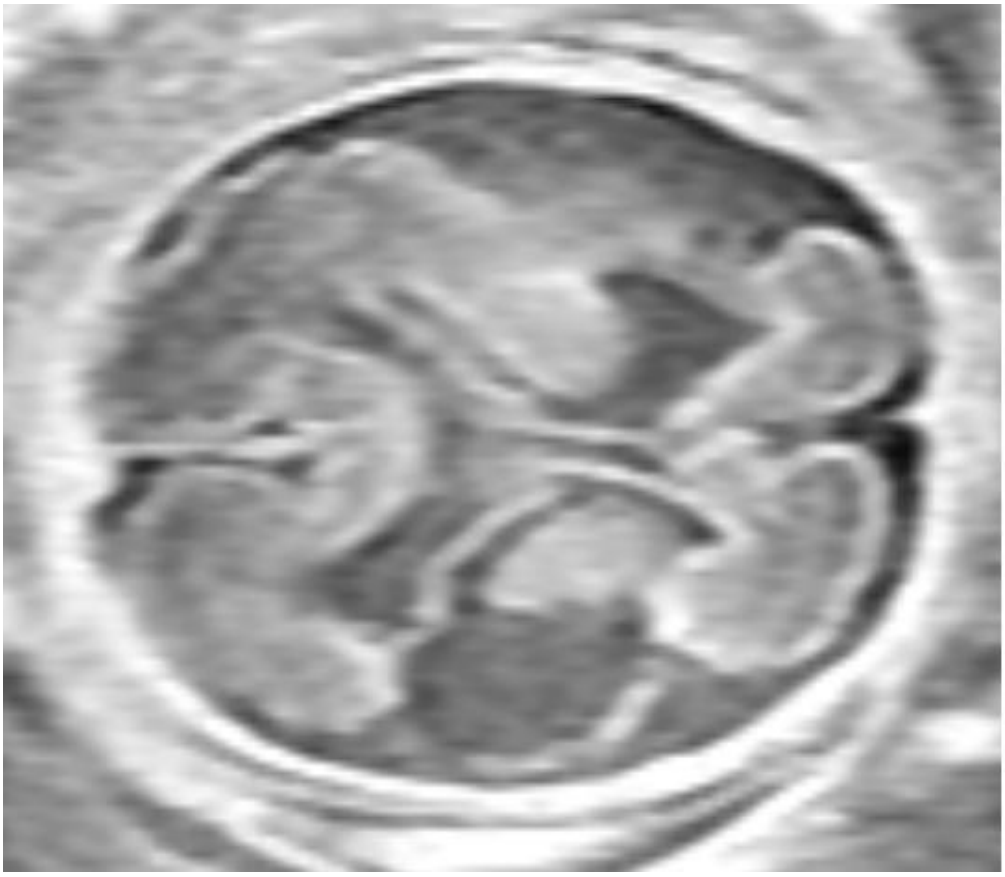
Figure 3

a



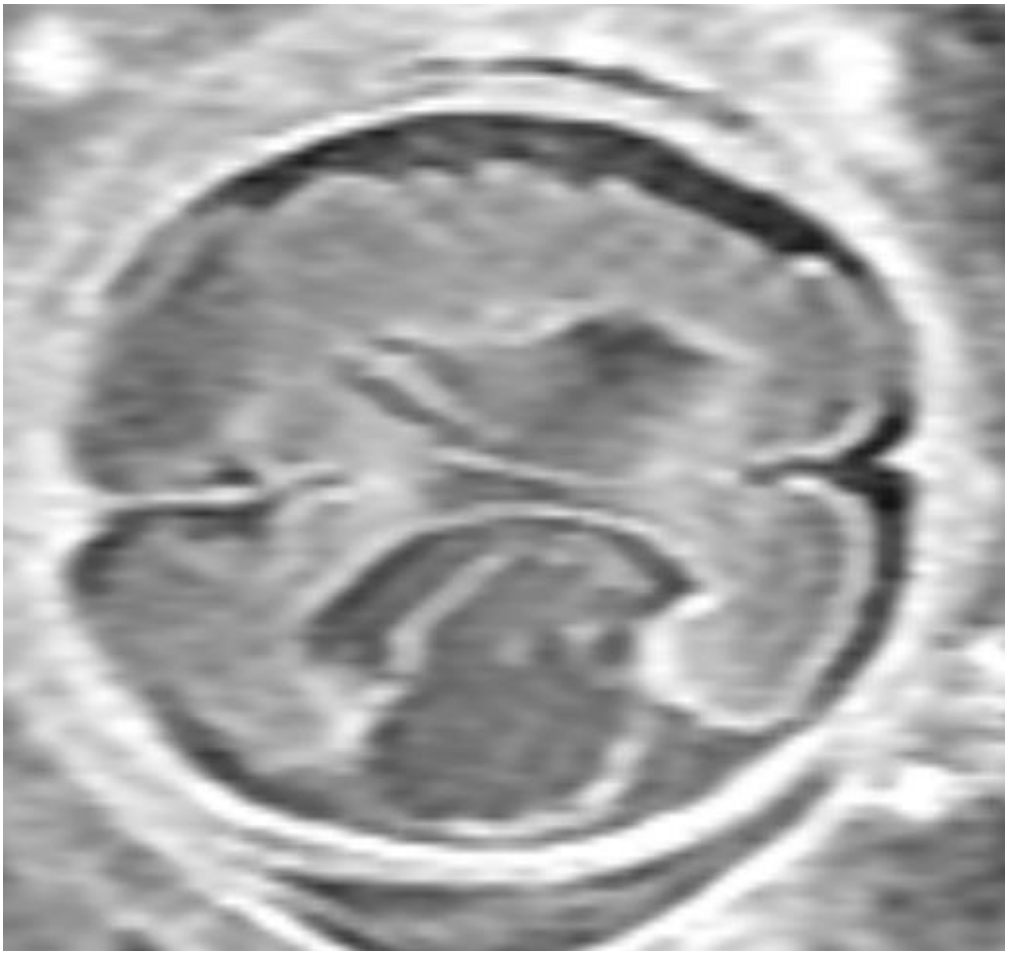
Description: Fetal brain shows bilateral open lip schizencephaly with asymmetry of the lateral ventricles at the schizencephaly defects. **Origin:** Augusta University

b



Description: Fetal brain shows bilateral open lip schizencephaly with asymmetry of the lateral ventricles at the schizencephaly defects. **Origin:** Augusta University

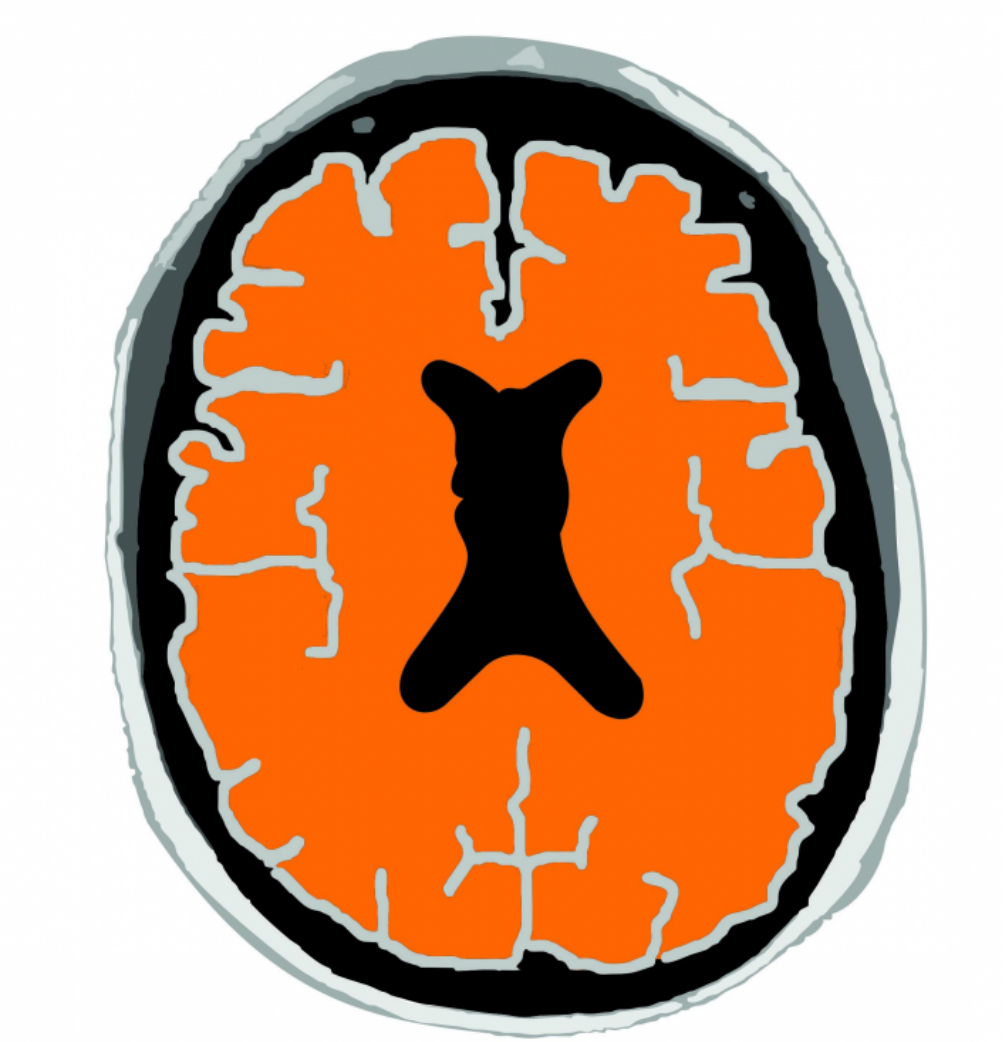
c



Description: Fetal brain shows bilateral open lip schizencephaly with asymmetry of the lateral ventricles at the schizencephaly defects. **Origin:** Augusta University

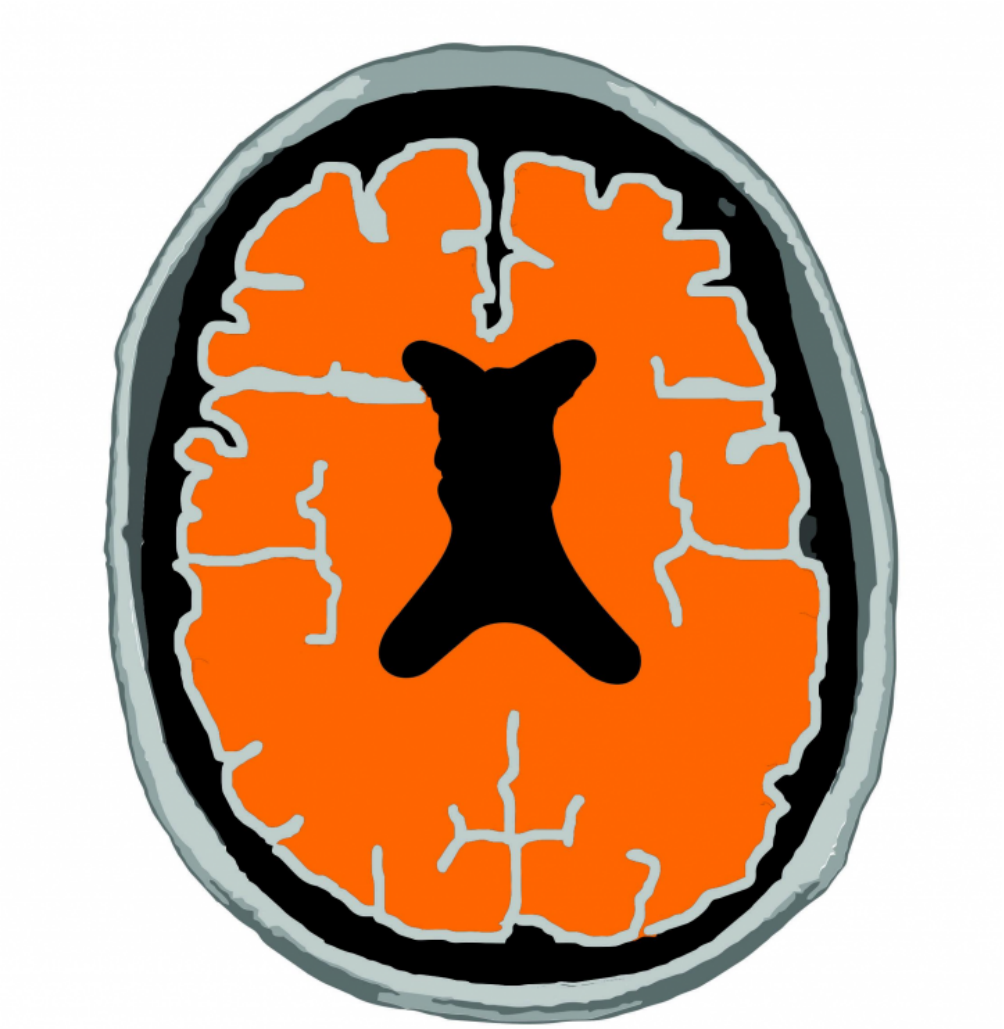
Figure 4

a



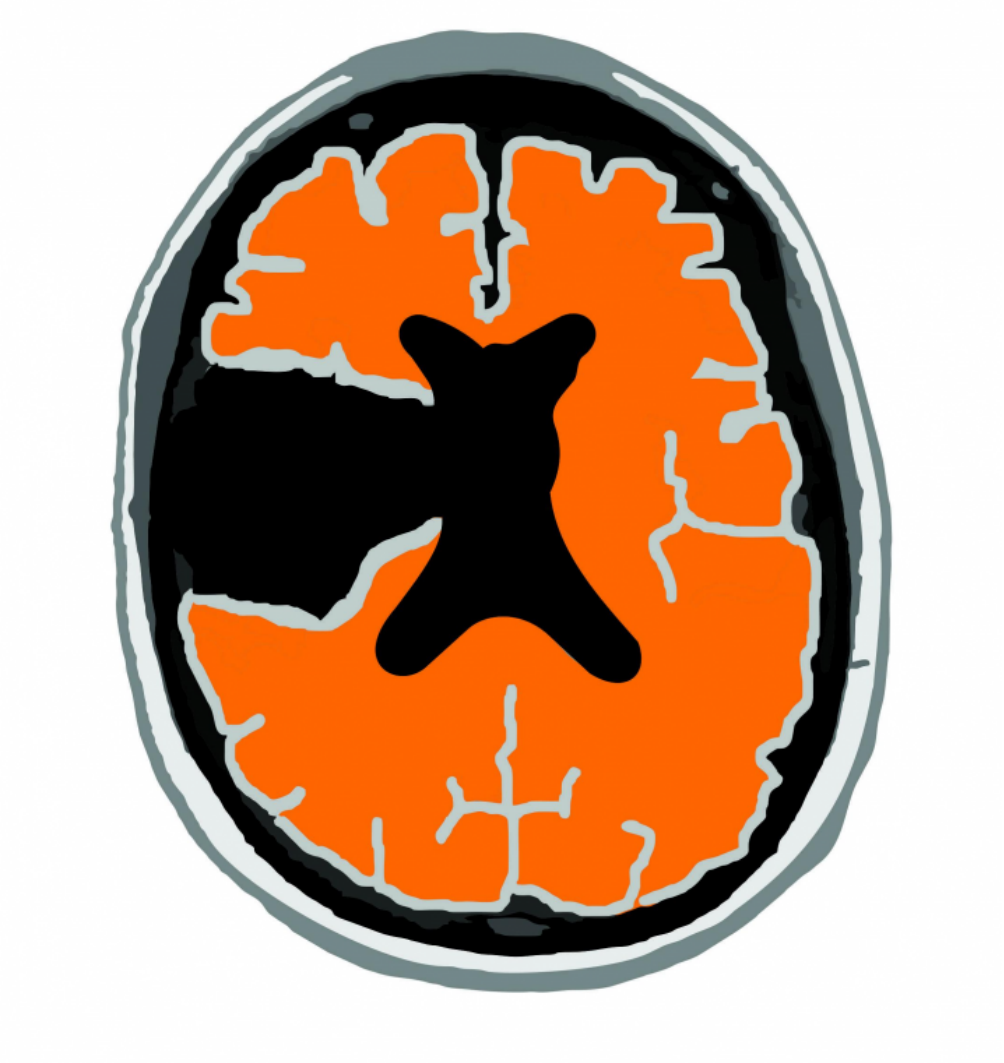
Description: Normal brain MRI **Origin:** Augusta University

b



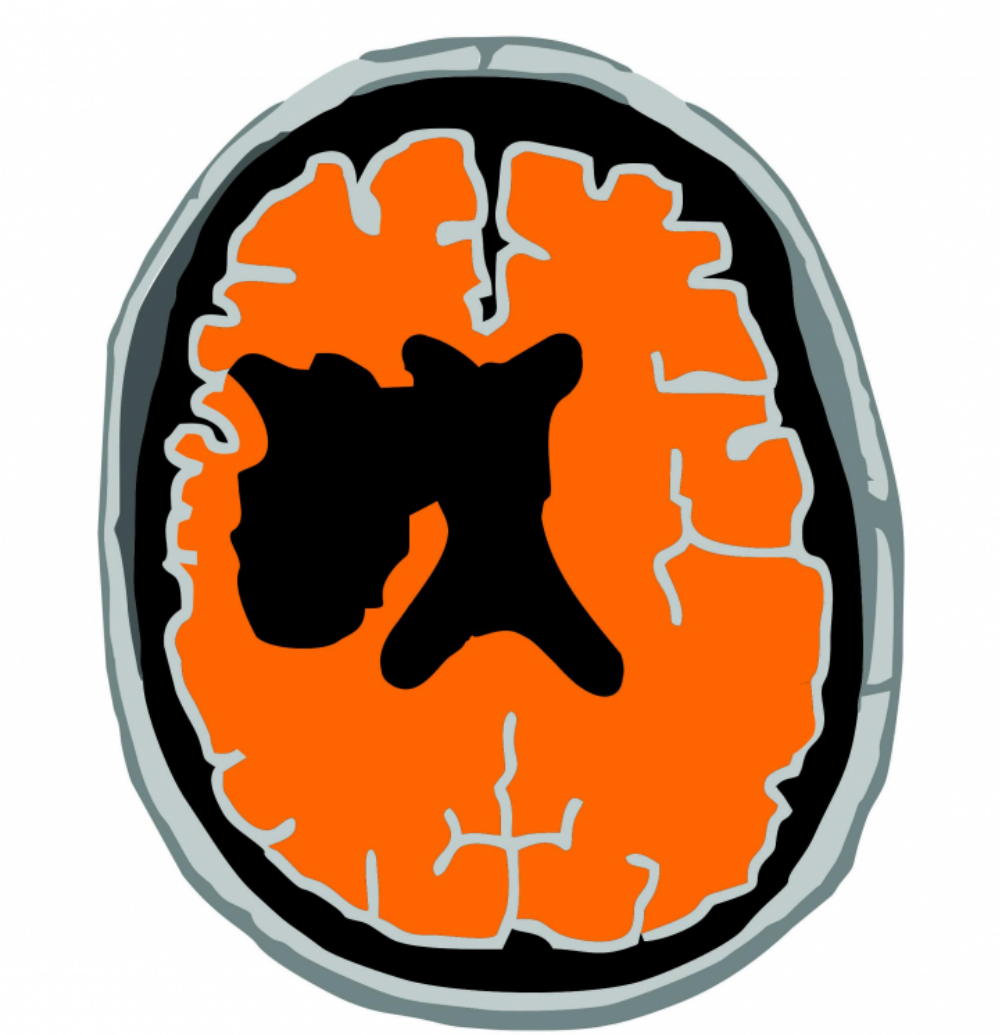
Description: Close-lip schizencephaly (type I). Notice the narrow cleft lined by dysplastic cortical grey matter. **Origin:** Augusta University

c



Description: Open-lip schizencephaly (type II). Notice the wide cleft from cortical surface to the ependymal margin, lined by dysplastic cortical grey matter. **Origin:** Augusta University

d



Description: Porencephalic cyst. Notice that the cyst opens to the ventricular space, expands into the periventricular white matter and is not lined by cortex but by gliotic tissue. **Origin:** Augusta University