



FREAKS OF NATURE

Foetal Monstrosity-**INIENCEPHALY**:A CASE REPORT

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Introduction: Foetal monstrosities are rare, iniencephaly, a neural tube defect is rarest, first described by Saint Hilaire in 1836. Probably due to complex alteration of embryonic development (defective mesoderm) during 3rd or 4th week of intrauterine life.

Classified by	Type 1	Type 2
1.Lewis	Iniencephaly clausus-no encephalocele	Iniencephaly apertus – with encephalocele
2.Howkins & Lawrie	Simple iniencephaly	Iniencephaly with anencephaly

Incidence is 0.1 to 10/10,000 live births

More common in female, (M:F- 1:9)

Chance of recurrence is 1-5%.

Case report:

History:

21 years, primi gravida, 20 weeks gestation

low socio economic status, unbooked and unimmunised

no history of folic acid or other drug intake

non consanguineous marriage

no family history of neural tube defects or congenital abnormality, no addictions

Diagnosed as simple iniencephaly clausus with cardiac abnormality, medical

termination of pregnancy done using vaginal misoprostol. Post-mortem Chromosomal analysis not done. Findings of the abortus (female) and fetogram were consistent with the ultrasound findings.

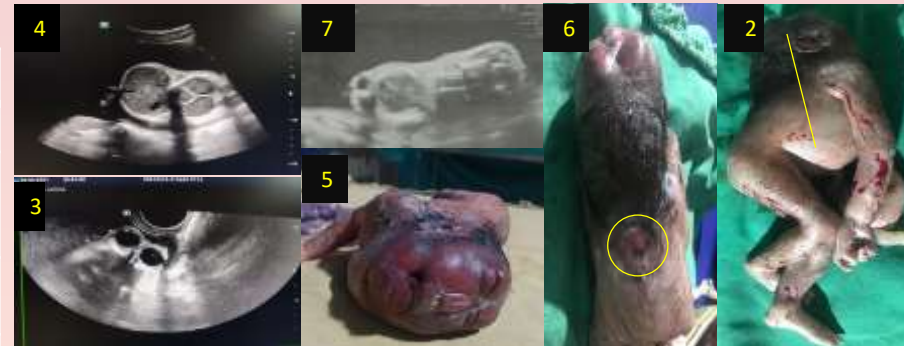
Discussion: Iniencephaly is derived from Greek word inion which refers to back of the neck and encephalos which refers to the brain.¹ The fusion of posterior most part of occipital bone with back leading to the absence of the neck and retroflexion of head.² Exact aetiology and pathology is not known, genetic and environmental causes may have a role.¹

	Ultrasound findings - 20 weeks fetus
1	short rotated spine with rachischisis
2	fixed retroflexed head with extreme hyperextension at neck, deficit of occipital bone
3	star gazing appearance, both orbits appear protruding, with hypotelorism
4	hypoplastic right ventricle and left ventricular outflow tract abnormality
5	cleft lip with protruding tongue
6	lumbar meningocele of size 2.4x2.2 cm
7	Abnormal intracranial anatomy, abdomen and head in same line
8	Normal long bones
9	polyhydramnios

Risk factors: Low socio economic status, low parity, deficiency of folic acid, obesity, hyperhomocysteinemia, and drugs, sulphonamide, tetracycline, antihistamines, chlomiphene citrate and anti tumor agents.¹ Associated chromosomal abnormalities: trisomy 18,13, monosomy X.¹

Differential diagnosis: Anencephaly with retroflexion of spine, Klippel-Fiel syndrome, cervical meningomyelocele, spondylocostal dysostosis, nuchal tumors.¹

Prognosis: It is always fatal in the neonatal period, four cases with mild iniencephaly clausus have been reported with long-term survival, with minimal morbidity.⁷



Conclusion: Iniencephaly is a rare occurrence, and diagnosing it in the early gestational period is critical, so as to offer medical termination of pregnancy, as almost all cases are invariably fatal and if survival is established, morbidity is severe. This is an attempt to make the mind know iniencephaly, so that our eyes picks it up and helps us treat the same.

Reference:

1. Sherif Elsirgany, Samesh Salama, Mahmoud Al Alfy and mona Aboughar. A rare case of a fetal neural tube defect: Iniencephaly. Imaging Med (2018) 10 (1)
2. Pungavkar SA, Sainani NI, Karnik AS et al. Antenatal diagnosis of iniencephaly: Sonographic and MR correlation: A case report. Korean. J. Radiol. 8, 351-355 (2004)
3. Chen CP. Prenatal diagnosis of iniencephaly. Taiwan. J. Obstet. Gynecol. 46, 199-207 (2007)
4. Aradhya Himabindu, Kanaka Durga Prasad Bhamidipaty, et al, Iniencephaly associated with anencephaly – a rare neural tube defect. Journal of critical reviews, ISSN 2394-5125 Vol 7, issue 11, 2020.
5. Saint-Hilaire IG. Iniencephalus. In: Bailliere JB, ed. Histoire des Anomalies de l'Organisation, Vol2. Paris, 1836;308-10
6. Lewis HL. Iniencephalus. Am J Obstet 1897;35:11-53
7. Katz VL, Aylsworth AS, Albright SG: Iniencephaly is not uniformly fatal. Prenat. Diagn. 9, 595-599 (1989)
8. Ballantyne, J.W. Antenatal pathology and Hygiene