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FREAKS OF NATURE-INIENCEPHALY

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.

Type 1: Iniencephaly clausus-no encephalocele

Type 2: Iniencephaly apertus – with encephalocele

other classification- with anencephaly and without anencephaly

Incidence is 0.1 to 10/10,000 live births, M:F- 1:9, Chance of recurrence is 1-5%.

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.

Ultrasound findings: short rotated spine with rachischisis

fixed retroflexed head with extreme hyperextension at neck, deficit of occipital bone, star gazing appearance, both orbits appear protruding, with hypotelorism

hypoplastic right ventricle and left ventricular outflow tract abnormality

cleft lip with protruding tongue, lumbar meningocele of size 2.4x2.2 cm

Abnormal intracranial anatomy, abdomen and head in same line.

Diagnosed as simple iniencephaly clausus with cardiac abnormality, medical termination of pregnancy done. Post-mortem Chromosomal analysis not done. Findings of abortus were consistent with the ultrasonogram and fetogram.

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

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 pick it up and help us treat the same

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