CYCLOPIA

(A Case Report)

by

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Introduction

The international incidence as cited by W.H.O. for congenital malformed babies ranges from 0.44% to 0.59%. Although the commonest malformation noted was of the central nervous system the occurance of cyclopia has not been mentioned. In Karnatak Medical College, Hubli from July 1976 to September 1977 the incidence of congenital malformations was 3.1%. Of these, there was 1 case of cyclopia which is reported not only due to its rarity but because of the paucity of literature on this anomaly.

Case report

P. Aged 36 years gravida 3 was admitted for elective repeat lower segment caesarean section on 16-9-1977. In her 10 years of married life her previous 2 deliveries were both by lower segment caesarean sections, the first one for breech 8 years ago and the second for uterine Inertia 2 years back. Both the babies were normal. The first male baby expired due to infective hepatities on the 4th day and the second is a female alive and healthy. It was very unfortunate that in this third section, the baby was a monster which survived for 4 hours. Placenta on examination showed no abnormality.

On further enquiry, the patient gave history of no medication, and was positive that she was careful in consuming any drugs. There was also no history of infection or irradiation.

The patient had a smooth postoperative period and was discharged on 26-9-1977.

Description of the Monster: (See photo 1)

I. **External features**

Weight 2.9 Kg.
Sex. Female.
Crown head length: 41 cms.
Circumference of skull 25 cms.
Face: was abnormal.

The forehead was narrow with a central diamond shaped fissure and a centrally placed fused eyeball. The eyelids were rudimentary with small eyelashes.

(b) The nose was absent. Instead there was imperforated proboscis located above the median eye 2.5 cms. long.

(c) On either side of the median eye, faint eyebrows were visualised.

(d) The mouth was small—2cms (microstomia). The philtrum was absent.

(e) The palate was closed and placed high.

(f) Both ears were normal.

The rest of the body was normal except the hands and soles of the feet had only a single crease.

II. **Radiological description of skull**

(a) **Face:** There was fusion of the orbital processes of the zygonatic and frontal bones forming a single orbit at the centre. The ethmoid, inferior turbinate, nasal and lecrimate bones were absent. The palate was closed and high arched.

(b)Anterior and posterior cranial fossae were deficient but midcranial fossa was well developed.

(c) **Vault:** was abnormal. Frontal suture was absent.

III. **Autopsy findings**

Scalp hairs were normal, although the skull was small. The major part of the base of the skull was occupied by the middlecranial fossa, the anterior fossa being situated in the middle portion of the skull and disproportionately

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small. The entire brain weighed 15 grams and was cystic. The cerebral hemispheres were fused with absence of convolutions, and no falx cerebri. Cerebellum was also small. Carpus call sun, septum plucidum and fornix were absent Olfactory bulbs were absent and only one optic nerve was seen.

All the other viscera were normal.

Comments

Cyclopia is commonly encountered in pigs and very rarely in human beings. It is formed in the early somite embryo i.e. 28 to 42 days after ferilisation. In this very early stage of embryogenesis, there is failure of development of frontonasal process and brain. The metabolic rate is reduced at a particular peak of division or differentiation of cells altering the development especially when the notochord is closing, Cyclopia involves the entire proencephalon.

The various factors in causing this defect in embryogenesis at this very early stage may be mostly due to hypoxia caused by infections, narcotics or respiratory depressants. The exact aetiology is still not identified at this early stage of pregnancy, as noted by Sainaba *et al* in 1972 in the single case reported from Calicut.

Summary

A single case of Cyclopia is reported.

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See Fig. on Art Paper V