CYCLOPIA

(A Case Report)

by

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Cyclopia is one of the rarest congenital malformations seen in human beings. In this there is a single centrally placed orbital fossa with the eye balls fused completely or partially. Other malformations may be associated. Edmonds (1950) has reported a series of 53 cases and Connaughton (1970) 3 cases. We came across only one case in a period of 6 years.

CASE REPORT

A 40 years old multigravida was admitted for confinement in a nursing home at Bhagalpur. She had 5 normal deliveries with all the children living, having no congenital abnormality. There was no history of congenital malformation in the family or any drug intake or infection or irradiation during early pregnancy. The labour was prolonged and features of obstructed labour developed. There was foetal tachycardia too. Lower segment caesarean section was done under general anaesthesia as an emergency procedure. The malformed baby was asphyxiated and was a typical cyclops with hydrocephallus. The baby died immediately though respiratory efforts were made by the baby.

Description of the Cyclops

It mas a male baby weighing 2.1 kg with gross hydrocephalus. Ears were placed at the angle of jaw. The mouth was represented by an opening 0.5 cms in diameter with small lips. The eyes were placed centraly with central cornea separating the pupillary areas. The eyelids with eyebrows of both sides were set like a rhomboid. There was a nose bud with a stump which originated 0.5 cms above the superior angle of the rhombus. There was complete non-canalization of the rudimentary nose. Skull bones were thin and the hair was well developed. Frontal bones were very small and parietal bones were relatively big with central bony deficiency. The two parietal and occipital bones were separated by a big fontanelle measuring 14 cms longitudinally and 11 cms transversely.

Post Mortem findings

On opening the fontanelle, haemorrhagic cerebrospinal fluid drained out. The total amount of the cerebrospinal fluid was approximately 540 ml. The brain was compressed and underdeveloped though it had all it's components.

The maxilla and mandible were not separated. The testes, urethra and anus were normal. Bladder had urine in it. Liver, spleen, gall bladder, kidneys, intestines, heart and lungs were normal. Floatation test for lungs was negative. The oesophagus was patent.

Measurements

Crown-rump length 55 cms. Circumference of head 45 cms. Circumference of chest at the level of nipples 22.5 cms. Circumference of abdomen at the level of umbilicus 20.5 cms. Length of neck 3.75 cms. Length of each upper limb 17.5 cms. Length of each lower limb 17.5 cms.

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Discussion

Cyclopia is more commonly seen in animals. In human beings it is an extremely rare congenital malformation. This consists of a single orbital fossa with one eye or two fused eye balls. A single rhomboid shaped palpebral fissure is present as in the present case. The eyelids are very small or at times may be absent. The nose is either completely absent or represented by a tubular structure situated above the centrally placed eyes, as in this case (Fig. 1). It has no communication with the pharynx. The brain always shows abnormality. The cerebral hemispheres are usually fused in the anterior region or may be fused throughout. The brain mass gets reduced to 1/3rd its normal volume. The parts of brain below the cerebral hemispheres and spinal cord are usually normal. The occulomotor and abducent nerves are either absent or hypoplastic. The pituitary gland may be absent. In Edmond's series of 53 cases (1950) 6 had absence of pituitary gland. According to Howorth *et al* (1961) chromosomal aberrations are usually not associated with cyclopia. However, no study in this regard was carried out in our case due to lack of facility.

The exact aetiology is not known. Stockard (1909) had produced such anomaly experimentally. He demonstrated that by temporarily reducing the metabolic rate at a time when a group of cells are at the peak of multiplication subsequent development of that group of cells might be profoundly altered. If the reduction in metabolic rate occurs as the neural tube closes, cyclopia occurs, Connaughton and Dew (1970) suspected it to be a drug induced abnormality. However in the present case no such history was available. Cases reported by Joshi (1974) and Sainaba *et al* (1972) had associated Hydramnios too which is an uncommon association in cyclopia.

Summary

A case of cyclopia with gross hydrocephalus and other associated congenital malformations has been presented.

References

1. Connaughton, J. and Dew, E.: Brit. J. Cl. Uract. 24: 37, 1970.
2. Edmonds, H. W.: Arch. Path. 50: 727, 1950.
3. Howorth, J. C., Medovy, H. and Lewis, A. J.: J. Pediat. 727, 1961.
4. Joshi. L.: J. Obst. & Gynec. India. 24: 89, 1974.
5. Sainaba. M. K., Indira, O. and Nayar, V.: J. Obst. & Gynec. India. 22: 87, 1972.
6. Stockard 1909 quoted by reference, 1.

See Fig. on Art Paper V